Discussion

If it is assumed, as is indeed probable, that the "cold" reported in Case I was a haemolytic streptococcal infection, then in each of the cases the rheumatic manifestations were preceded by such an infection. Another feature of all four cases is the absence of a previous history of rheumatic infection and of any family history of this disease. On the other hand, there is much variation in the period intervening between the onset of the haemolytic streptococcal invasion and that of the rheumatic manifestations. In view of the clinical picture and the response of these cases to sodium salicylate, there can be little doubt that the cardiac and arthritic signs were of rheumatic origin.

It must be borne in mind, however, that this is not a random selection of cases. For every case here reported there were many more where either haemolytic streptococcal infection was present without associated rheumatism, or acute rheumatism existed without any evidence of haemolytic streptococci being involved, although the former was the more common finding. In other words, haemolytic streptococcal infection without any accompanying or consequent rheumatic manifestations was much more frequently found than acute rheumatism without some evidence of haemolytic streptococcal involvement. This is a factor which is of the greatest importance in any attempt to assess the role of the haemolytic streptococcus in the aetiology of acute rheumatism. It has been pointed out elsewhere¹¹ that if any series of cases of haemolytic streptococcal infection be examined the majority of cases of acute rheumatism in the series will be found to be secondary to such infection, whereas in a series of cases of acute rheumatism the number of cases secondary to haemolytic streptococcal infection will be much smaller.

The cases here reported are deliberately chosen because of the dominant part played by the haemolytic streptococcus, and in order to emphasize the important, though perhaps not primary, role of this organism in the aetiology of acute rheumatism, a role which must always be considered in any attempt to investigate the aetiology of this disease. That a close association does exist between the haemolytic streptococcus and acute rheumatism is exemplified even in this short series, where the latter condition was found in association with: (1) tonsillitis, (2) otitis media, (3) mastoiditis, (4) puerperal fever, (5) pyosalpinx, (6) acute nephritis, (7) acute osteomyelitis, (8) erysipelas, (9) impetigo, and (10) scarlet fever, in each case haemolytic streptococci being isolated as the causal organism. To argue from this, however, that the micro-organism is the primary cause of acute rheumatism is not valid, in view of the much larger number of cases in which no evidence of acute rheumatism is ever found.

The value of the cases in this series is further enhanced by reason of the fact that in none was there any family or previous history of acute rheumatism; it is not possible to argue, therefore, that we are here dealing with the invasion of tissues already susceptible to the disease. Again, the period elapsing between the first signs of haemolytic streptococcal invasion and the onset of the rheumatic manifestations is so variable that it argues strongly against the haemolytic streptococcus being the primary cause of the disease.

As suggested elsewhere,¹¹ the present position would seem to be one of two alternatives: either that infection with the haemolytic streptococcus facilitates the invasion of the tissues by some specific agent, or that the haemolytic streptococcus may so alter the tissues susceptible to rheumatic infection as to prepare the way for invasion by this specific infective agent.

I have to acknowledge my great indebtedness to Lieut.-Colonel John Cunningham, superintendent of the Astley Ainslie Institution, not only for permission to study the cases under his charge and for full access to the case notes, but also for the sympathetic assistance which I received from him at all times. I am similarly indebted to Professor Charles McNeil for permission to publish the notes on Case 1. This work was part of a larger study of acute rheumatism which was being carried out in co-operation with Dr. H. J. Gibson of the department of bacteriology, and before my entrance to the Royal Navy, with the aid of personal and expenses grants from the Medical Research Council, and latterly during my tenure of the Davidson Research Fellowship in Bacteriology.

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MENTAL DEFICIENCY AND HEREDITY

BY

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A little over a year ago I completed an investigation into the incidence and circumstances of retarded school children and mental defectives, which had for one of its objects the investigation of medical and family histories of all defectives of school age within a defined area. An attempt was made to estimate the relative importance of various factors in the causation of mental deficiency, and it is the purpose of this paper to give a brief account of this aspect of the original investigation. Other relevant matters are referred to from time to time, as appears necessary to the purpose in view. If in places this short contribution seems ragged, and poor in supporting evidence, indulgence is asked on the ground that it is the essence of a thesis some ten times its length, an important part of which was the presentation of genealogical charts which consideration of space precludes from reproduction here.

Introductory

The area referred to is the north-east quadrant of East Suffolk, with a total population of 45,435, a school population of 6,645, and a density of population of 0.3 persons per acre. The chief industries are agriculture and fishing. Communications across country are relatively poor, and many villages are still comparatively isolated. Social cohesiveness is high throughout the area, and remarkably complete family histories were obtained, as a rule, without great difficulty. The standards employed were those fully described by Dr. Lewis in his report.¹ A total of 750 children (11.2 per cent. of the entire school population) was examined by individual methods, and there is every reason to believe that the ascertainment of defectives was, for practical purposes, complete. Stanford tests and Burt's graded tests were used respectively in the estimation of mental and of educational age. The investigation of suspected defectives of school age followed closely that usually undertaken in the completion of Form 306 M_a of the Board of Education; that of suspected adults was made with the social concept of deficiency in mind. It was neither practicable nor indeed desirable to subject adults to systematic testing.

While it is clearly impracticable to submit any details of individual cases, it may be said generally that judge-

ments relating to school children were based on precise observed facts, those relating to parents on personal observations, and those relating to other members of a family on strongly presumptive evidence, corroborated whenever possible.

Total Ascertainment and Family Histories

A total of 165 ascertained defectives of school age was made up of 135 feeble-minded, twenty-three imbeciles, and seven idiots, a proportion almost identical with that found by Dr. Lewis in rural areas.² The close correspondence is a matter of importance, because it raises a presumption that no great disparity existed between the standards employed in the investigations.

The incidence of mental defect and retardation ran hand in hand, and the essential feature of loci of high incidence of both was their degree of social isolation. There was no ground for believing that this factor operated directly, but it was, in all probability, the biggest cause of migration from such areas to others of greater social amenity, a movement most affecting the better stocks of higher initiative. Consequently, the proportion of inferior stocks in these areas was increased, and the incidence of deficiency rose *pari passu*.

A total number of 116 family histories was investigated, twenty-eight of which were not full enough to permit conclusions being drawn with any confidence. No fewer than 149 of the ascertained defectives of school age were involved in the family histories, leaving only sixteen, all of feeble-minded grade, in whose cases a family history was unobtainable. It is convenient here to present summary pathological and aetiological classifications based on the medical and family histories.

Pathological Types			
Imbeciles and Idiots		Feeble-minded	
Mongolians ...	9	Infantile encephalitis ...	2
Meningitic ...	1	Traumatic (birth injury) ...	1
Hydrocephalic ...	1	Simple primary ...	132
Epileptic ...	1		
Simple primary ...	18		
Total ...	30	Total ...	135

Causation			
Imbeciles and Idiots		Feeble-minded	
(a) Environmental factors	6	(a) Environmental factors	4
(b) Without apparent cause	17	(b) Without apparent cause	10
(c) Prima facie evidence of a hereditary factor	7	(c) Neuropathic inheritance	71
		(d) Combination of neuropathic inheritance and environmental factor	15
23 per cent. attributable to inheritance.		71 per cent. attributable exclusively to neuropathic inheritance.	

No further comment need be made on Group (a), and it will be sufficient to say of Group (b) that cases under this heading could not be referred to a cause, although reasonably complete information was available. It is likely that a fuller knowledge would have led to their classification with those due to environmental causes.

It is necessary now to indicate the nature of the evidence which was taken into account in the selection of cases attributed to neuropathic inheritance, either alone or combined with an environmental factor. Very briefly, inheritance was regarded as a factor in causation in the presence of: (a) several defective siblings; (b) defectives or other marked neuropaths in antecedent generations, especially defectives or borderline parents. Without elaborate statistical methods, themselves not beyond criticism by statisticians, conclusive proof of hereditary influence cannot be offered, but it is noteworthy that other workers who have used the method of family surveys have reached substantially the same conclusions as those presented here. A glance at the family charts referred to would give a very fair notion of the standard of judgement employed, but a personal acquaintance with the families in their usual environmental setting is by far the most convincing evidence. It is hoped that the following numerical summaries will give some idea of the features of the family charts of feeble-minded children whose amentia was attributed to neuropathic inheritance.

Number of defective sibs (a) in family	1	2	3	4	5	6	6+
Number of families ...	19	4	7	5	1	—	—

Total (b) size of sibship ...	1	2	3	4	5	6	6+
Number of families ...	5	8	8	9	4	—	2

Number of unsatisfactory (c) siblings	58	= 79 per cent.
Total number (b) of siblings	124	
Number of defective or borderline (d) parents	40	= 60 per cent.
Total number (b) of parents	66	

(a) "Sibs" means siblings in family of an ascertained defective of school age. (b) Total number does not include those who were not assessed either because they were inaccessible, or in the case of children too young. (c) "Unsatisfactory" means defectives, borderline cases (I.Q. less than 80 per cent.), dead during infancy, and stillborn. (d) "Borderline" means illiterate dullards who were almost certainly defective in an educational sense during childhood.

Feeble-mindedness was judged to be due to the combined influence of hereditary and environmental factors where, in the presence of a neuropathic family history, only one of many siblings was actually defective, whereas several others were of poor mental quality, although not actually defective. It seemed that a factor, sometimes suggested by the history, additional to the neuropathic inheritance had operated to produce an actual defective among a group of poor quality siblings.

Lower-grade Deficiency and Heredity

It will have been remarked that in 86 per cent. of the feeble-minded there is evidence of hereditary influence alone or in combination, in contrast with a *prima facie* 23 per cent. of lower-grade aments. Dr. Lewis, who reached a similar conclusion after a more extensive investigation, says: "Lower-grade deficiency is not a family problem to the extent that some previous writers have led us to expect." If all grades of defectives are considered together 75 per cent. appear to be associated with a neuropathic inheritance. As the incomplete histories, which all refer to feeble-minded persons, are not taken into account in this estimate, it follows that the proportion of cases in which hereditary influence plays a part is somewhat underestimated, for an undue proportion of lower-grade defectives is admitted.

Dr. A. F. Tredgold, referring to defectives of all ages, types, and grades, estimates that in approximately 80 per cent. the condition is due to inheritance.⁴ He points out that inquiries which are confined to children will show a relatively greater number due to environment, because lower-grade cases, in which the expectation of life is considerably less than in higher-grade cases, are due, for the most part, to environmental causes. Bearing in mind the omission of many incomplete family trees of higher-grade defectives, the restriction of the initial ascertainment to children, and the relatively small number of cases, the present estimate appears to be in close agreement with that of Dr. Tredgold.

It is important to realize that the majority of feeble-minded children are merely profound dullards, distinguished from those usually classed as "dull and backward" by a purely arbitrary borderline. Such deficiency has no pathology in the ordinary sense of the word, and the subjects thereof are represented by the extreme left of a frequency distribution curve of intelligence quotients. They are quite properly regarded as the analogues of short individuals of say five feet, in a community of average height 5 feet 6 inches, in which a few members attain heights of 6 feet. It is among this group and not among definite pathological types that hereditary influence is so prominent.

According to my own investigations twelve out of thirteen children certifiable as feeble-minded according to educational standards are well behaved during the years of school life, and exhibit no distinctive abnormalities of character other than apathy and undue docility.

Furthermore, roughly half the children certifiable as defectives for the purpose of the Education Acts will not be defective in a social sense in adult life in a rural area.⁵ That is to say, there exists a wide disparity between the standards used in the educational and social classifications of feeble-mindedness, and it is difficult to avoid the conclusion that the use of the single term "feeble-minded" in two distinct senses, both of which are legal and not scientific, is highly confusing in an inquiry of this sort. An attempt was made to overcome the difficulties implicit in this duality of meaning, but finally a conviction was established that nothing short of a carefully planned genealogical survey, extending over at least two generations, would meet the case. Be that as it may, a study of families containing defectives enabled certain principles, which are summarized below, to be advanced tentatively, yet with the knowledge that they are consistent with, and lend support to, many widely known authoritative opinions.

Summary of Conclusions

1. Roughly 75 per cent. of defectives of all grades come of stocks which exhibit distinct mental abnormalities, of which deficiency and gross dullness are the commonest.

2. Of the remaining 25 per cent., a few are attributable to environmental causes, but most are without apparent cause.

3. Feeble-mindedness and dullness are familial to a much greater extent than lower-grade deficiency.

4. The mating of two mentally defective individuals yields offspring who are all defective.

5. The mating of two individuals of "poor type" yields offspring of "poor type," who are often defectives in an educational and sometimes in a social sense.

6. "Dull parents, dull children," is a usual association.

7. When a neuropathic stock exists the chance presence of an adverse environment, using the term in its broadest sense, may produce actual deficiency in cases which might not have been defective without the intervention of such a factor.

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Clinical Memoranda

PERFORATIVE APPENDICITIS COMPLICATING ACUTE NEPHRITIS IN AN INFANT

Acute appendicitis under 1 year is very rare, and under 6 months excessively so. Of 500 cases admitted to the Royal Infirmary, Edinburgh, there were but three in infants in the first year of life (Fraser¹: he does not give the exact ages in months). Pre-natal appendicitis is recorded by Jackson²; Kümmell, quoted by Lipshutz,³ records appendicitis in a baby who died twenty-four hours after birth. Abt⁴ has made what is probably the most exhaustive survey of the literature, and has collected records of seventy cases in children under 2 years, twenty being in infants under 3 months. Reports by other observers covering a large series of cases show a similar striking paucity of references to appendicitis in infants less than a year old. Fraser⁵ gives an anatomical and bacteriological explanation for this. (1) There is a very small amount of lymphoid tissue in the appendix at birth, and it gradually increases; the presence of this tissue seems to render the locality in which it resides more prone to infection. (2) The *B. coli communis*, which is the infecting organism in the majority of cases, shows great variation in virulence; just after birth it is very slightly toxic, but this feature becomes more marked as the child grows older, probably caused by change in diet and in some cases by attacks of gastro-enteritis, mild or otherwise. The following case is therefore of interest.

A female, aged 5 months, was admitted to hospital on December 5th, 1933, suffering from general oedema. We could ascertain nothing beyond a history of a rash six weeks previously; there had been measles in the family, and this probably accounted for the rash. (Appendicitis has been quoted as a complication of measles by some observers, and Thenebe, Hirshberg, and Cenci⁶ have reported six cases, but in none of them was the patient under 5 years; it is doubtful, of course, if there is any real connexion in this case.) The urine was scanty and loaded with albumin, and a diagnosis of nephritis was made. On December 7th no urine was passed, and on the following day 1½ c.cm. salyrgan was given, which produced a copious flow. Urine was passed freely on December 9th, but was scanty again the following day. The salyrgan was repeated on December 11th, and the flow again became copious. On December 12th the child passed urine freely and appeared much better; on December 13th and 14th the urine was freely passed, but salyrgan was repeated on the 14th. The child died on December 15th.

The temperature was 99° F. on admission, 100° on the sixth day, and rose to 104° on the seventh; it continued between 102° and 103° until death, when it rose to 105°. The stools were green and slimy on admission, but became normal after three days until the last two days, when only mucus was passed. Owing to the difficulty of obtaining a specimen of urine no record was made of the amount passed. The usual methods of producing diuresis—hot packs, etc.—had no apparent effect. The abdomen was examined on a few occasions, but beyond being oedematous—and this was improving—there was nothing else to be noted; the child slept fairly well, and took its feeds.

At necropsy the kidneys showed the features of a nephritis; they were not enlarged, but were congested, cortex thinned, and there were spots of haemorrhage scattered through the medulla. Beyond some slight congestion of the lungs the other organs were normal. On looking into the pelvis pus was discovered, and on further investigation the appendix was found to be perforated at the base; the pus which had gravitated anterior to the rectum was odourless, and there were no obvious signs of peritonitis. The brain was not examined, as the parents did not wish the child's head or face to be disfigured.

There is no record, so far as I am aware (but this is subject to correction), of appendicitis complicating acute nephritis in an infant. In this case it was an absolutely silent appendix: Lipshutz remarks that infants with appendicitis always vomit, but this feature was absent; the infantile peritoneum is said to be very susceptible to infection, and a general peritonitis rapidly develops, but this did not occur. Which condition killed the infant—the nephritis or the appendix? And am I correct in assuming that the rise of temperature marked the entrance of the appendix to the picture? The case is interesting, and I should like the comments of those more experienced than myself.

I am indebted to Mr. E. S. Gawne, F.R.C.S.Ed., medical superintendent, for permission to publish this case.

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