A NEW WHITE-LOCK FAMILY (BARTON'S CASE).

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The hereditary nature of the "flare"—or pigmentless forehead patch accompanied by a white or yellow white-lock of hair—in man is now well known. Several illustrative pedigrees were published by Nettleship, Usher and Pearson in their Monograph on Albinism*, and a good pedigree was also published by Dr E. A. Cockayne in 1914 under the title of "A Piebald Family †." That family embraced six generations of the hereditary flare. In the present case we have only been able to trace four generations with a total of seven affected members. Dr Cockayne's pedigree involved nineteen affected members, while Rizzoli's pedigree of the Bianconcini family extends to six generations with twenty affected members. I owe the case to Dr E. R. Barton, of University College Hospital, who came across III. 1 and IV. 10 in the Midwifery Department of the hospital and most courteously put me in touch with the family. The pedigree should accordingly be referred to under Dr Barton's name.

There is no consanguinity. The family goes grey early and there has been a certain amount of tuberculosis.

I

II

III

IV

No consanguinity.  * Died at birth

I. 1. T. H. Normal, died at about 40.
I. 2. J. F. Married I. 1 and is the earliest known ascendant to have the flare. She had both white forehead patch and white lock. She died at about 80.

† Biometrika, Vol. x. pp. 197—200, with eight plates.
II. 1. R. H. White forehead patch and white lock. She married but has had no children.

II. 3. E. H. Married L. L. (II. 4). E. H. died at about 40 and had white forehead patch and white lock; not known whether he had any body patches.

II. 5. J. H. Normal, married II. 6, normal. She has had five normal children. One of these, III. 15, died at 25, the other four III. 7, 9, 11 and 13, all married and had again normal families, IV. 11, 12, 13 and 14.

II. 7. H. H. Normal, married a normal woman, II. 8, and had a boy and girl, III. 16 and 17, both normal.

II. 9. A. H. Normal, married a normal woman, II. 10, and has had a normal son, III. 18.

II. 11 and 12 died young, but are said to have been normal.

The family of II. 3 consisted of three daughters and two sons of whom only one was affected.

III. 1. R. J. H. Has a yellow-white lock and white forehead patch, said to have no white body patches. She married III. 2, W. S. R., a normal.

III. 3. A. W. H. Died owing to an accident at 24—25 years. He was normal.

III. 4. L. H. Was normal and died at 8 years, of meningitis.

III. 5. Normal, died as a baby.

III. 6. D. H. Normal, is alive at 28 years and unmarried. Thus the trait could only be carried on through III. 1. She has so far had only ten children, but three of them are affected.

IV. 1. R. R. Now 18 years old, is normal.

IV. 2. L. C. R. Died at 15½ years. She had the white forehead patch, the white lock and white patches on the skin.

IV. 3. W. R. Aged 14, and IV. 4, G. R., aged 11½, are both normal.

IV. 5. A. R. Aged 9, has yellow-white lock, white forehead patch, but no skin patches elsewhere.

IV. 6. D. R. Aged 7½, and IV. 7, G. R., aged 5, are normal.

IV. 8. Died at birth, and nothing is known of her.

IV. 9. B. R. Aged 1½, is normal.

IV. 10. T. C. R. Aged four weeks when seen, has quite a marked forehead patch and white lock on his fairly profuse dark hair. No white body patches.

There are three marriages—non-consanguineous—of affected with apparent normals, each of which produce affected. The affected in each case produce affected. The character therefore cannot be recessive. We are compelled to treat it as dominant, but as I. 2 has some normal offspring, she must have been heterozygous. It will be clear that none of the affected can be looked upon as pure dominants, and in their families there ought to be 50% affected.

We must leave out II. 11 and II. 12 as there is no record, and IV. 8 who died at birth although III. 1 considered the infant normal. We have accordingly 2 in 5,
Barton's White-Lock Family
1 in 5 and 3 in 9 in generations II., III. and IV. affected or 6 in 19. This is a fair approximation to a third, but rather a poor one to a half. There is in fact a deviation of 3.5 with a standard deviation of $\sqrt{19 \times \frac{1}{5} \times \frac{1}{5}} = 2.179$, or the deviation is 1.61 times the s.d. The odds against such a defect are more than 17.5 to 1. Of course not impossible but improbable.

On the other hand there is no case in which an apparent normal marrying a normal has transmitted the character. In the case of IV. 11, 12, 13 and 14 there are considerable families, exact numbers not available, but it is known that none are affected. Thus it would appear that transmission through the unaffected either does not ever, or at least commonly, occur.

This rule holds also for Cockayne’s family and for the Bianconcini.

IV. 2 is the only one with body pigmentless patches. As III. 1 mentioned this fact without special enquiry, I think we must take it that her statement with regard to IV. 5 and IV. 10 and to herself that they and she have no pigmentless body patches can be accepted. She believed that II. 1 had no such patches, but did not know about II. 3 or I. 2. It would seem therefore that in this family at least conspicuous body patches are not frequent. It should not therefore be looked upon as a piebald family in the ordinary sense. It exhibits the inheritance of a “flare.”