

Records of Y-inherited hairy ears in India

by
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This striking condition has been regarded as extremely rare. Indeed our knowledge of its inheritance is based on a single family in Italy (Tommasi 1907) in which there were eleven cases, all males, in five generations. (See pedigree in Gates (1946) p. 13, and p. 249). The inheritance was only from the father to all his sons, and to all their male descendants. It must therefore be determined in the Y-chromosome. Even this has been doubted, because the family was described nearly fifty years ago, when methods of recording pedigrees were undeveloped. In this Italian family the condition was associated with alcoholism and insanity, an association which will be discussed later.

While traveling with my wife in Uganda last year I encountered a case of hairy ears in the captain of a steam launch which sails at intervals between Butiaba on Lake Albert and Murchison Falls on the Victoria Nile. It was recognized at once by the long, black hairs rising obliquely from the rim of the ear, which I was kindly allowed to photograph. (See Figs. 1 and 2). While essentially the same as the Italian case, certain differences in hair distribution will be discussed later. The Captain was an East Indian, indeed a Goan, from Goa, the Portuguese enclave on the West coast of India. His father died when he was very young, so he knows nothing of his father or grandfather. But his two brothers both have hairy ears, while his three sisters do not. He has four sons, whom I interviewed afterwards with their mother in another part of Uganda. As the eldest is only twelve, none of them had yet begun to develop hair on the ears. The father said that his ears began to show the condition when he was 22 years old. These and other data are thrown together in pedigree form in Fig. 3. While consistent with Y-chromosome inheritance, this family would not by itself show the method of inheritance.

On more careful examination of the Captain's ears, the long hairs were observed to arise somewhat in tufts from the ear margin only. Mainly they originated in the groove between helix and antihelix, but a few were from the cartilage of the ear rim itself. They were black, like the scalp hair, but less wavy. Although soft and pliable they stood out stiffly from the ear, projecting somewhat upwards (Fig. 2) and forming a loose, black fringe which is visible at some distance. He refuses to have them cut lest it make them stiff. In this case and in two other Indians to be described below the earlobe was devoid of hairs. This condition of the Captain's Ears was referred to a novel by Hemingway.

The next case was encountered by my wife in a bank at Fort Portal. He was

Fig. 1

a Sikh, living in Uganda but born in India in the district Kochiakur. As is well known, Sikhs wear a turban and never cut their hair, so at the age of 45 he had a full beard and mustache. The hairy ears, are therefore less conspicuous, but the distribution of the hairs was found on examination to be exactly the same as in the previous case. He had previously been in the Indian army and was a man of fine physique. His photograph is not included here because the hairy ears are not very clear in contrast with his hirsute face. Questions elicited the fact that his father and his two brothers all had the same hairy ears. His son is only twelve years old, so again the facts are compatible with Y-inheritance but would not by themselves prove it.

The third man was another Goan. He was a steward on the P. & O. ship to Marseilles, which we boarded at Port Said. He had the same distribution of hair, in the ear rims but not on the lobes. His father had this condition, but he was uncertain whether it is present in his three brothers.



Fig. 2

Discussion

The fact that representatives of three independent families with hairy ears of the same type, all from India and two of them from Goa, were discovered casually while traveling in Africa, seems to indicate that the condition is by no means infrequent in India, and especially in Goa. In all three the hair distribution in the ears was the same, and the presumptive method of inheritance was holandric. Moreover, the 8 individuals interviewed in this connection were all of normal mentality and there was no hint of mental aberration in any of their relatives.

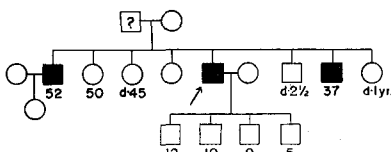


Fig. 3 - Pedigree of hairy ears.
The figures represent ages

That hairy ears are not uncommon in Western India is supported by casual observations of Mr. David L. De Harport while he was photographing the Ajanta frescoes in the northwestern part of Hyderabad. He reports seeing that the head photographer of the Department of Archeology in Aurangabad, which had recently moved from Poona, had notably hairy ears. One of the waiters in the hotel at Aurangabad where Mr. De Harport lived for several months wore a turban but also had hairy ears. These places are 200-300 miles northwest of Goa and inland from Bombay.

The growth of hairs in the external auditory meatus of older men is of course not unusual, but their appearance in the rim of the ear at the early age of twenty years or less is an entirely independent phenomenon. Tommasi (1907) cites several other cases in the Italian literature, to which I may now refer. At that time the work of Lombroso had stimulated an active interest in a possible relationship between ear characters on the one hand and criminality, insanity, alcoholism or epilepsy on the other. The earliest of these papers cited, (Chiarugi 1889), refers to Darwin's tubercle and also describes hairs, often pigmented in the fetus and in young babies, on the ear margin in two streams which meet at the position of Darwin's tubercle. Gradenigo (1890) in general terms discusses ear forms in relation to crime and psychiatry. The view that such a relation exists was widely held in Italy at the time, but Gradenigo confines himself to ear form and mentions no case of hairy ears. The following three authors, however, describe specific cases of hairy ears in Italy, and in two of these at least the condition is the same as described by Tommasi.

Cainer (1898) described two brothers with this condition, hypertrichosis of the ears. Photographs of the two ears of the elder brother, who was 87, show the same condition as the photographs of Tommasi (1907). Besides the peripheral long outstanding hairs the medial portion of the ear was also bushy with hair. The Italian

cases thus showed more extreme hypertrichosis than the three East Indians I have observed. This may indicate a different gene, but it may perhaps be only an age difference. The Italians photographed were older men, Tommasi's case being photographed at 55 (but he was still living at 84 years of age) and Cainer's case apparently at 87, whereas the Indians I saw were all young to middle-aged men, the Sikh (45 years) being the oldest. The two brothers described by Cainer were both asylum inmates and recorded as insane. The younger brother, who was 75, had been convicted at various times and had attempted suicide. He was an epileptic who finally became insane.

In the Tommasi pedigree (Gates 1946, p. 13) II, the paternal grandfather of the proband (III.5) was an alcoholic. His father (II.3) was an unbridled alcoholic who died intoxicated, having fallen and fractured his skull. His mother (II.4) was addicted to wine and spirits and died of apoplexy. His brother (III.6) was a strange character whose daughter (IV.1) died in childbirth. The proponent himself (III.5) was also addicted to alcohol. He was first in the Bologna asylum at 42 but was dismissed in four months with the recommendation to abstain from alcohol. At 52 he was returned to the asylum but dismissed in 165 days. However, he was back again at 55 and spent his old age in confinement. He was first married at 26 and later married again, but had no children. His sister (III.7) was also mental. As his mother was a drinker and his sister a mental case, there is no reason to suppose that the gene for hypertrichosis of the ears was pleiotropic, affecting both ears and mentality, or had any connection with the mentality except as a *possible* instance of linkage between the gene for hairy ears and a gene affecting the mentality. As the former gene is in the Y-chromosome and the proband's sister was mental, linkage was not involved, even assuming that a gene affecting the mentality was present in the family. It may be added that Tommasi measured the length and breadth of the head and of the ears of the proband, but the derived cephalic index and the ear index are both given incorrectly.

Batistelli (1899) observed further cases of hairy ears. He says he has seen the presence of long hairs in the ear border and on the face, *especially* (my italics) in criminals, epileptics and idiots. In the case of an idiot described the hairs were erect, dense, rigid, 4 cm. in length, covering the anterior surface of the ear. This is undoubtedly the same condition as described by Tommasi and by Cainer. Batistelli concludes that hairs are more frequent around the organs of sense and the genitals in « degenerates » than in normals, but much more evidence would be required to support such a conclusion.

Pianetta (1901) describes another case of hypertrichosis of somewhat different character in a man of psychopathic type. The hairiness of the ears appears to be the same as in the other Italian cases, but it is accompanied by hairiness of the body, and the ear is also misshapen. The ears were rather small and not the proper « ovoid » shape. There was no Darwin tubercle and the lobe was small and adherent. There is nothing remarkable about this (see Gates (1954) but the ears were provided with hairs « recalling those of pithecoïds ». The helix had no distinct fold, but was narrow and thin, adhering to the rest of the ear so there was no scaphoid fossa. The

earlobe was covered with fine hairs; much more robust hairs were implanted on the border of the ear and perpendicular to it. There was also a tuft of hairs in the meatus. The long border hairs were evidently the same condition we have been considering. But in addition there was an abundant development of hair covering the trunk down to the waist. The hairs though rather fine, were long and thick everywhere, including the shoulders, where they continued on to the arms, including the deltoid region. On the back especially they were at some points wavy and curly. The hair on the stomach was normal, and there was no sacral or coccygeal hypertrichosis, but more hair was found on the calves than on the anterior of the legs. The beard was heavy, covering also the zygoma.¹

This man was in an asylum, suffering from hallucinations and exaggerated emotive states. He was epileptoid and finally showed mental disintegration. He was regarded as a case of (atavistic) hypertrichosis combined with hereditary psychopathic degeneration.

From the above evidence, it would appear that this particular type of hypertrichosis of the ears may be as common in Italian as in E. Indian males. All the cases observed in Italy have been in connection with mental defect. At that time, through the influence of Lombroso, « stigmata of degeneration » were sought for in the ears of the insane and of criminals. It would therefore be unsafe to conclude, without a modern study of the Italian population, that there is any necessary connection between hypertrichosis of the ears and any mental defect. Such a study would probably reveal a much higher frequency of the gene for hairy ears than has been assumed, and any association with insanity or other mental aberrations might prove to be purely incidental.

Summary

While traveling in East Africa, three typical cases of hairy ears were observed incidentally in natives of India, two of whom were from Goa. The inheritance in all three cases was compatible with location of the gene in the Y-chromosome. Two other independent cases have been reported to me from the same general part of India, southeast of Bombay, which indicates a considerable frequency of the gene in this area.

The only previously known pedigree of this condition came from Italy in 1907, and several independent cases in males are described in the early Italian literature. In all these Italian families the hairy ears were associated with insanity or other psychiatric conditions, but this association may have been fortuitous, and hairy ears may be as frequent in Italy as in India.

¹ A hairy body is characteristic of the Ainu of Hokkaido, but in the personal examination of 140 adult Ainu of both sexes in 1954 no case of hypertrichosis of the ears was observed.

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RIASSUNTO

Tre casi tipici di orecchie pelose sono stati osservati per caso, durante un viaggio in Africa Orientale, in individui nativi dell'India, di cui due di Goa. L'eredità di tutti e tre i casi era compatibile con una localizzazione del gene nel cromosoma Y. Due

altri casi indipendenti mi sono stati riferiti dalla stessa area dell'India, a sud-est di Bombay il che sta ad indicare una considerevole frequenza del gene in tale area.

L'unico caso famigliare precedentemente nato di questa anomalia veniva dall'Italia nel 1907, e diversi casi indipendenti in

maschi sono descritti nella più lontana bibliografia italiana. In tutte queste famiglie italiane le orecchie pelose erano associate a pazzia od altre malattie psichiatriche, ma tale associazione potrebbe essere stata fortuita, e le orecchie pelose potrebbero essere altrettanto frequenti in Italia che in India.