CASE III

Male, aged 53, suffered from a sudden attack of auricular fibrillation in September, 1927. He came under the care of Professor T. G. Moorhead, who has kindly permitted me to record his case. During a period of eighteen months after his initial attack of irregularity, repeated attacks occurred of paroxysmal fibrillation with some flutter. The heart showed moderate enlargement, with a systolic murmur maximal in the mitral area. Complete examination failed to show any cause for the irregularity. His teeth were carefully examined by a dentist in November, 1928, and were pronounced sound. Early in 1929 the patient began to suffer from some discomfort with his teeth, and a general deterioration of health was noted, as evidenced by considerable loss of weight, with increasingly frequent incidence of fibrillation. In April, 1929, he consulted a London cardiologist. His teeth were x-rayed and two roots were condemned. Removal of all his teeth under local anaesthesia was recommended. In May, after an abortive attempt to improve the condition of pyorrhoea which then existed, two teeth were removed without difficulty, the only anaesthetic employed being cocaine, which was simply painted on the gums. On June 27th another tooth was removed. This extraction proved difficult, and some novocain was injected into the gums. Following the extraction the gums continued to bleed for three days. The bleeding ceased after application of adrenaline. On July 1st a purpuric eruption occurred all over the body. On the next day there was pronounced haematuria, and oozing of blood was noted all over the surface of the palate and from the gums. Haemostatic serum and antistreptococcal serum were administered and the bleeding ceased. Four days later there was haemorrhage from the stomach and bowel, and, from this on, the patient was mildly pyrexial. Blood culture was negative. At this time I saw the patient in consultation on two occasions. No marked change was noted until August 14th, when he was apparently much better and able to go for a drive. On the next day he was not so well, and was again feverish. Haematuria once again appeared, and there were more purpuric spots. Two days later it was noticed that the left arm was paralysed. Half an hour later the left leg was paralysed, and still later the left side of the face. Towards evening there was severe haematemesis, after which the patient sank into coma and died.

COMMENTARY

It will be seen that in two of the cases recorded above there was no symptom or sign of endocarditis until the removal of septic foci. The first patient had experienced no disability whatever from his heart lesion, and consulted a dentist because of trouble with his teeth. The second patient was led to consult a cardiologist because of rejection for the Army. His valve lesion had not interfered with indulgence in strenuous exercise. The third patient had presented a number of symptoms before the removal of teeth, but the terminal illness was closely associated with an extraction. I feel justified in including his case in this paper in spite of negative blood culture, as the circumstances associated with his death point the same moral as the other cases.

Many interesting problems arise from consideration of these cases. If proof were needed that infection in subacute endocarditis may derive from teeth or tonsils, such cases would supply the necessary evidence. Paradoxically, then, they emphasize the importance of removal of septic foci, but they stress the need for great caution. Removal of more than one or two teeth at a sitting in patients with disease of the heart is clearly unwise. Unless there is a definite periapical abscess it would seem equally unwise to risk dissemination of infection by removing teeth which are very firmly fixed and require much manipulation. General anaesthesia should on a *priori* grounds be preferred to the use of local anaesthesia. Preliminary vaccination with an autogenous growth from

the gums or tonsils seems a rational procedure. Careful supervision of teeth in patients with disease of the heart should obviate the necessity for wholesale extractions. Crowning of teeth in such patients and the preservation of dead teeth are both to be condemned. It is better to sacrifice a comparatively useful tooth at a stage when its removal is unattended with danger, than to preserve it in a condition in which it is likely to become severely infected and a source of danger whether it be left or removed.

THREE GENERATIONS OF INHERITED DENTAL DEFECT

BY

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Congenital absence of certain teeth is a relatively common condition which sometimes occurs in family groups, but gross suppression of the majority of the teeth in members of three consecutive generations of the same family is rare.

Recently a woman attended an infant welfare centre with her only grandson, a boy 8 months old, who had six teeth already erupted. Mention of these teeth led the woman to volunteer the statement that her own two widely spaced teeth were the only ones she ever possessed. These peg-shaped teeth, occupying the place of the lower central incisors, appeared when she was about the age of 7, and are still strong and sound, being implanted in firm gums, which, she says, have always been capable of dealing unaided with all kinds of food except nuts. During childhood, complete closure of the mouth was impossible owing to hypertrophy of the gums, but it was not until she reached the age of 19 that, goaded by her companions' constant teasing on account of her toothlessness, she sought advice at the local hospital. Two "levelling" operations ("for double odontoma," she says) took place, after which the gums shrank to their present normal proportions, and she was able to close her mouth naturally.

Her memory regarding the results of x-ray examination then and later is vague; but apparently when she was 19 a few rudimentary teeth were said to be present, and again, twenty-seven years later, when she was in another hospital for gynaecological treatment "almost a full set of teeth" was found. Operation on the mouth was then suggested, but she would not agree to this as her bite was satisfactory, and her enunciation clear and unusually

good. She is one of a family of four, all the others having normal dentition. Her age is 46, but her appearance is that of one twenty years or so older. She is stout and florid, and her abundant dark hair is going grey. "A complication of disorders"—the graphic phrase of some former biographers—might sum up her past and present health, for she has never been strong, having suffered from "chronic bronchitis, gall-stones, appendicitis, and displaced kidney," and having had, eight years ago, after several curettages, a major gynaecological operation, of the nature of which she is unaware.

THE FIRST GENERATION

The mother (Generation I) of the above patient, who died at an advanced age, was one of a family of eleven children, and was "the only one with bad sight or anything the matter with her teeth." She had "no teeth until she was grown up, and then two came right under her tongue." She was short-sighted, a good speaker, and photographs show the usual edentulous sinking-in of the lower part of the face such as occurs after multiple extractions or in old age. History farther back is unobtainable, except that the family, on each side, has been country-born and bred for many generations, and has always been used to an ample diet containing fresh milk and vegetables. Despite this woman's appearance, no Semitic element is known on either side of the family, and the living members now under consideration are of good intelligence and physique.

THE THIRD GENERATION

The third generation consists of the only grandson of the old couple, a young man, father of the infant first mentioned. At the age of 15 he had no erupted teeth, but now, at the age of 23 years, he is teething, and has at the present date two more teeth than has his 10-months-old son. His gums are still enormously broad, but evidently are much reduced following an operation done when he was 15. At that time localized pain and inflammation round the second upper right molar induced him to go to the dental hospital, where a local abscess was evacuated. X-ray examination, when he was 9, and in a children's hospital, "as he was not very strong," is said to have shown "few teeth in the jaws, and these only stumps," but six years later a full set of unerupted teeth was found to be *in situ*. Soon after this he was referred to a general hospital, where, after another x-ray examination which corroborated the above, he says both gum and bone were "chiselled off" and four teeth set free. Reference to an early photograph shows the inability to close the mouth prior to the operation, as happened in his mother's case; no such disability was present afterwards.

The right upper molar referred to juts out on the outer aspect of the maxilla at an angle of 35 degrees or so, and around it inflammation occurs occasionally, but to a less extent than before. The visible teeth are odd and asymmetrical in shape, though not at all Hutchinsonian; they are very broad proportionately, probably because they are only partly erupted, though strong and fairly firmly set. As far as their oddity allows descriptive enumeration, all the incisors seem to have erupted except the left upper one; the right and left lower canines are present, as is the left upper one. Behind the canines the gum in the lower jaw is less thick than in the upper jaw. The patient's enunciation and bite are now excellent, and he is an expert whistler.

A COMMON EYE DEFECT

Common to this family there is an eye defect to which, curiously, they have given little attention or treatment. The baby's grandmother has always had extremely defective sight, and has been almost blind for the last four or five years, dense bilateral corneal opacity being now present. She says she "brought this into the world with her," and apart from some treatment at the age of 9 or 10 years, its course has been unchecked. A year ago she was prevailed on to attend hospital, and was then told the damage was irreparable. She is active and handy in her house, though she leaves machining and hand-sewing to her son; she peers—though she can see neither the printed page nor a human face—closely at all she handles, declaring herself benefited by the use of strong

concave glasses purchased from a hawker for a relative now deceased. Her own mother, possessor of the "two teeth under her tongue," had, she says, "exactly the same kind of eyes as mine," with very defective, untreated sight; and her father, a venerable old man by his photographs, always had poor sight, and went blind when he was 47. Both these old people had abundant hair. Her son, father of the baby in Generation IV, has a slight degree of bilateral keratitis with defective sight, for which he wears glasses, "fours or fives," obtained "from a young gentleman at the door" (who supplied the other glasses mentioned). He has neither had, nor desired, any other form of treatment. The infant, whose head and face show extreme asymmetry from whatever aspect they are regarded, has an alternating squint. He is well grown, but some developmental genital defect is also present, for neither testicle is descended, and the scrotum is absent, or almost so. He plays happily in his cot, and is already quickly responsive to the sound of a piano or band, but he is considered by his grandmother to be "not at all well," owing to teething.

COMMENTARY

The available bibliography illustrates the heritability of similar dental defects, either fixed from one generation to another, or variable; and also an association with certain epiblastic defects, not present in the family under consideration. This family differs also from Thurnam's case (*London Med. Chir. Soc. Trans.*, 1848, p. 71) of two cousins, each of whom had only four teeth and was the subject of congenital alopecia, the scalp hair being represented by lanugo only. He also quoted additional cases. Danz, according to Michelson (*Ziemssen's Handbook*), saw two adults who had never had either hair or teeth. Another condition of congenital absence of cutaneous appendages is known in which teeth, hair, and sweat glands are all either rudimentary or suppressed. Such subjects, with their undeveloped alveolar processes and normal development of the upper part of the face and head, suggest in their features a curious resemblance to a foetal skull, and are apt to become severely distressed in hot weather owing to heat retention, and inability to reduce their body temperature by sweating. A small family group of two persons who appear to correspond to the above type has been recorded by Dr. Jean Smith, in two successive communications in the *Proceedings of the Royal Society of Medicine* (1929, xxii, 649; January, 1931, xxiv, 323); two brothers showing this dental defect, along with absence of sweating and a tendency to attacks of pyrexia (apparently from insufficient heat loss in absence of sweating), and scanty hair. The special value of her records lies in the x-ray picture accompanying the second record, to which reference may advantageously be made. She alludes to Clouston's account in the *Journal of the Canadian Medical Association* (1929, xxi, 18) of a French-Canadian family in or near Quebec in which 119 of such cases are said to have occurred throughout six generations. Apart from this family group she states that she had only succeeded in tracing the record of fifteen similar cases in the literature. Thadani, however, reports (*Journ. Hered.*, 1921, xii, 87) a Hindu Amil community in India, in which a toothless type of man occurs. The men are not only toothless, but bald-headed and extremely sensitive to heat. The evidence, as far as it goes, indicates that the character is male sex-linked. Anodont females are not at present known, but if daughters of a toothless father are married to a normal man their male children are toothless, and they seem to be known locally as bhudas, or old men. Crossman (*Journ. Hered.*, 1927, xviii, 368) cites from Guildford in 1883 a not dissimilar type of case, in that of an edentulous woman without teeth or hair. She had a son in the same condition.

He married a normal, and of their children six were normal and two others had several undeveloped teeth. A normal sister had eighteen children, one of whom was without teeth or hair. Fürst (*Arch. f. Rass.- u. Gesells.-biol.*, 1905, xvi, 310) describes another case of anodontia.

Any possible connexion of rickets in the etiology of this condition need not be considered here, as a hereditary genetic defect displayed in three successive generations cannot possibly be due to such or to any other form of malnutrition or vitamin deficiency.

From all the foregoing cases this family group is essentially different, in that the primordia of the teeth are not suppressed, being present in the gums even when dentition is either greatly delayed or never completed. Sweating is normal here, and the hair is as abundant in all the affected members as it is in ordinary members of the general community. It is just conceivable that as the teeth are epiblastic appendages, and the corneal epithelium and the lens itself are also epiblastic in origin, the eye defect in this family and its inherited dental condition may be associated characters, joined or linked together in the process of heredity. Whether this is so or not must at present be left an open question, though the association itself is certainly suggestive.

I have to thank Dr. Veitch Clark, the medical officer of health, and Dr. Maude Duckworth, at whose infant welfare centre under the Manchester Corporation the infant was seen, for permission to publish notes of the above case; and to acknowledge to R. R. Gates's *Heredity in Man* (1929) my indebtedness for references to the literature.

HEREDITY AND DUPUYTREN'S CONTRACTION

BY

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The hardening and contraction of the palmar fascia, resulting in flexor contraction of the fingers named after Dupuytren, has been known for many years to have had a hereditary factor in its etiology. This factor has been thought usually to be of a gouty nature, influenced by occupation. It is rare to come across three brothers affected with the deformity, whose father was also affected, and whose mother was affected by double-bent little finger at the proximal interphalangeal joint, but whose other fingers were normal.

The father died at the age of 73, and the mother at the age of 63. All the affected followed different occupations. The father was a grocer, and the brothers were a baker, clerk, and general labourer, their present ages being 72, 70, and 68 respectively. There is no history of gout in the family. The pedigree is as follows (a female infant in Generation II, who died in infancy, is not shown):

Description of Members of Pedigree

I 1: Both hands affected; third and fourth fingers contracted; began at about the age of 50.

I 2: Both little fingers (fourth) bent at proximal interphalangeal joint; said to be from birth. Other fingers unaffected.

II 1, 4, 6: Unaffected.

II 2: Both hands affected; right first and fourth fingers, noticed first at the age of 57; left first, second, third, and

fourth, noticed first at the age of 37, after receiving injury to second finger.

II 3: Both hands affected; noticed first about the age of 40. Right first, second, and third fingers; left second and third fingers. Both fourth or little fingers are bent at the interphalangeal joints similar to his mother's.

II 5: Both hands affected; began at the age of 48. Right third and fourth fingers; left second, third, and fourth fingers.

III 6: Died in infancy.

III 7: Died, aged 23, unaffected.

III 1, 2, 3, 4, 5, 8, 9, 10: All unaffected—ages from 41 to 33.

It will be observed that there is some slight variation in the degree of deformity. II 2 and II 5 have the greatest deformity in the left hand, while in II 3 it is the right hand which is most affected. In all three the fourth or little finger is affected, and it is difficult to decide whether the maternal character as well as the paternal is shown, or whether the maternal factor has reinforced the paternal factor so as to produce the very marked deformity in all three brothers.

So far the deformity has not appeared in the third generation, but this generation has hardly yet reached the age when it begins to be noticed. The late period in life—advanced maturity—when the deformity appears and continues to progress, illustrates the potency of the hereditary factor which lies dormant during childhood, adolescence, and maturity.

There may be other defects or diseases appearing late in life which have a hereditary basis, but are not so easily recognizable clinically as Dupuytren's contraction. Owing to the difficulty of disentangling and analysing the various factors these defects or diseases are attributed chiefly to environmental causes.

Senescent hereditary defects or diseases are more difficult to study with the completeness of those occurring in earlier years, because many members of families do not live long enough for the defect to appear, and develop to such a degree as to be recognizable.

DISSEMINATED ENCEPHALO-MYELITIS FOLLOWING SPINAL ANAESTHESIA

BY

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Although spinal anaesthesia was introduced by Bier of Bonn in 1889 the method has been generally employed only since the discovery of stovaine in 1904. Toxic effects at the time when stovaine was on trial, and its known action in lowering the blood pressure and depressing the cardio-vascular and respiratory systems, discouraged many surgeons for a time from the employment of spinal anaesthesia. For many years it has been successfully used by those aware of its dangers and of the technique of administration; and in a large field of surgery it now appears to be the anaesthetic of choice. Those who employ this method have recorded series of thousands of cases without any immediate ill effects or sequelae, and the majority of deaths which have occurred under spinal anaesthesia have been due to the state of the patient prior to operation, shock, cardiac disease, and to loss of blood, etc.—conditions which would make the operation more hazardous if performed under chloroform or ether.

Headache, paraesthesiae, and even pain in the trunk and limbs, not uncommonly follow spinal anaesthesia. More rarely paralysis of an ocular muscle occurs, but recovery is usually complete within a few weeks. More