AN INHERITED JAW ANOMALY IN LONG-HAIRED DACHSHUNDS

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(With Plate IV and Six Text-figures)

Introduction

In this paper, we are going to describe briefly an inherited anomaly of the jaws which occurs very frequently amongst long-haired dachshunds. It appears that the same or a very similar anomaly is common in a variety of breeds of dogs. We therefore hope that it may be possible for other investigators to fill in some gaps which circumstances beyond our control have prevented us from closing.

Genetics

The condition to be described in this paper is very common. Practically every breeder of long-haired dachshunds in this country is familiar with it, though many will not admit that it has occurred in their own kennels. All long-haired dachshunds now kept in this country are derived from a small group of closely related animals which were imported from Germany after the Great War. An investigation of the pedigrees of affected animals shows that at least one, and probably several animals of the foundation stock must have been heterozygous for the condition. This consists of a bad fit of upper and lower jaw; the mandible is too short in proportion to the upper jaw, and consequently the occlusion of the front teeth is faulty; the animals are "overshot". Affected animals, though fully viable and vigorous, are valueless from the show point of view and are now rarely used for breeding. Practically all affected animals are therefore derived from two normal parents. As the data supplied by dog breeders are often unreliable, we have confined our material to all those litters which one of us (A. J. L.) has seen personally. The material is unselected, as it includes a large number of litters in which the condition did not appear. The ascertainment of affected animals within this group is therefore complete. The data are given in Table I, where r = number of recessives; s = size of litter; n_s = number of litters of size s; r_s = number of recessives in litters of size s; and t_s = total number of puppies in litters of size s.

Table I contains thirteen litters of normal parents which were not known to be heterozygotes, containing altogether seventy-four puppies. Of these, twenty animals were affected. As these data do not include

TABLE I

s	$r=1$	2	3	n_s	r_s	t_s
2	1	0	—	1	1	2
3	1	0	0	1	1	3
4	0	0	1	1	3	4
5	3	0	0	3	3	15
6	0	1	0	1	2	6
7	2	2	0	4	6	28
8	0	2	0	2	4	16
Total	7	5	1	13	20	74

those litters from heterozygotes which failed to contain an affected animal, a correction is necessary. The method has been given by Haldane (1938). The incidence of recessives (p) is derived from the formula

$$\frac{R}{p}=\sum_{s=2}^{S}\frac{t_s}{1-q^s},$$

where p is the frequency of recessives, and $p+q=1$; s is the number of puppies per litter; S = largest value of s in the sample; t_s = total number of puppies in litters of the size s; and R = total number of abnormals recorded. The formula gives p as the root of the equation

$$\frac{20}{p}=\frac{2}{1-q^2}+\frac{3}{1-q^3}+\frac{4}{1-q^4}+\frac{15}{1-q^5}+\frac{6}{1-q^6}+\frac{28}{1-q^7}+\frac{16}{1-q^8},$$

which is solved by a process of iteration. The result is

$$p=0{\cdot}1845\pm0{\cdot}0573,$$

the standard error being derived from the amount of information (I) concerning p according to the formula (Haldane, 1938)

$$I=\frac{R}{p^2q}-\frac{1}{q^2}\sum_{s=2}^{S}\frac{sq^st_s}{(1-q^s)^2}.$$

The value of p obtained is smaller than expected on a monofactorial basis (0·25), but the difference is not significant.

Two litters of four and six puppies respectively, were obtained from matings of two known heterozygotes. Each contained one affected animal. The ratio of eight normals to two abnormals is in agreement with a monofactorial expectation.

One litter of seven animals was obtained from an affected bitch by a dog known to be heterozygous. It contained four normal and three affected animals.

The data, therefore, leave no doubt that the anomaly is inherited as a simple Mendelian factor. It should be mentioned that the litter of two puppies in Table I contained one affected animal and one individual in which the condition was reversed, the animal being "undershot". As this anomaly has occurred but once, it is doubtful whether it has anything to do with the "overshot" condition described in this paper; the animal has accordingly been treated as a normal.

The distribution of the anomaly in the sexes is as follows. The animals in Table I included twenty-seven normal dogs, twenty-seven normal bitches, sixteen overshot dogs and four overshot bitches. Amongst the offspring of two known heterozygotes, there were three normal dogs, five normal bitches and two affected bitches. In the backcross litter, there were four normal dogs, one abnormal dog and two abnormal bitches. The sex ratio amongst the normals is undisturbed (thirty-four dogs, thirty-two bitches). Amongst the affected animals, there is an excess of males (seventeen dogs, eight bitches); the difference is, however, not significant ($\chi^2 = 3{\cdot}240$ for one degree of freedom; $P = 0{\cdot}072$). There is no indication of sex-linkage in the pedigrees.

MORPHOLOGY

The material available for the description of the anatomy and the development of the anomaly is very scanty, and many questions cannot be answered adequately.

We possess the skulls and atlases of a normal dachshund bitch, 14 months old, and of an abnormal dog of the same age; the animals are half-sibs (same father, two different heterozygous mothers). The dog had been buried for some time; the skull is slightly damaged across the left frontal bone, and the central upper incisors, the right upper third premolar and the left lower third molar are missing. In addition, we have kept a normal dog and an abnormal bitch (litter mates) for $8\frac{1}{2}$ months. Lateral view X-ray photographs of the heads were taken at the age of 47, 61, 89, 116 and 152 days, and X-ray photographs of the heads in top view were taken at the age of 89 and 152 days. A final inspection, particularly of their teeth, was made on the 261st day.

Four sets of differences may occur between the individuals. They are:

(*a*) Differences due to the main gene ("typical" differences).

(*b*) Differences due to other genes and the environment ("accidental" differences).

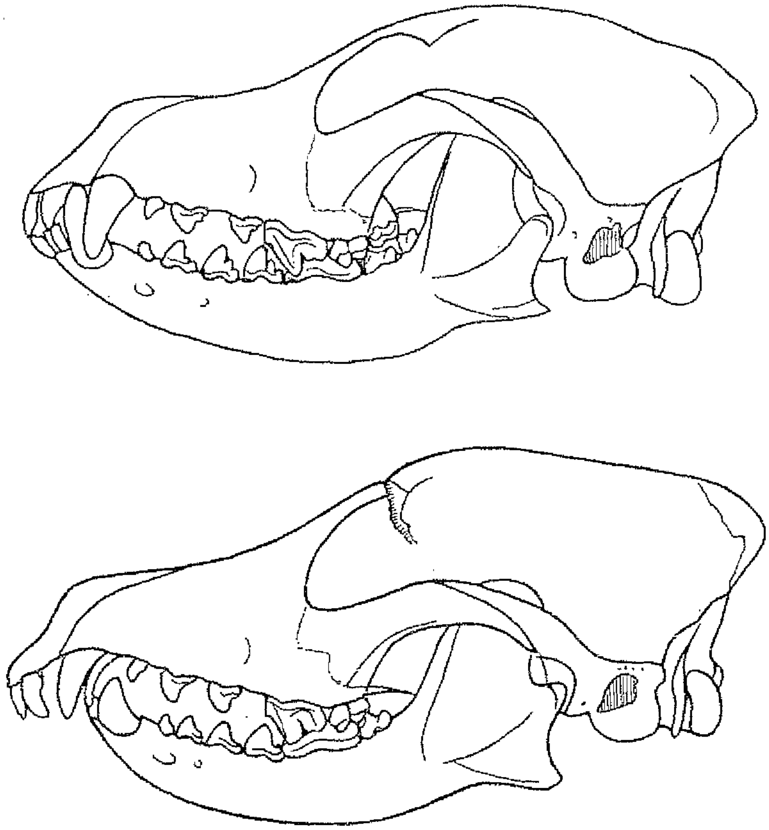
(*c*) Sex differences.

(*d*) Age differences.

These differences can be distinguished only in part. There is no age difference for the comparison between members of each pair of animals. As the last X-ray photos were taken when the second set of animals was nearly fully grown, at least so far as the skull is concerned, the age difference between the two sets of animals is practically negligible. Sex differences can be largely eliminated by the fact that the material contains both sexes of normals and abnormals. In practice it remains to distinguish between "typical" and "accidental" differences. Differences found between the two normals and between the two abnormals are almost certainly "accidental", whether genetic in nature or not. Resemblances between the abnormals which differentiate them from both the normals will mostly be "typical". The comparison is made more difficult by the fact that many features can be easily seen and measured on the skulls which are hidden in the radiographs. In view of these difficulties, we are giving extensive illustrations of the two skulls which are based on photographs (Text-figs. 1–6); many minor differences not mentioned in the text will be found there.

Living overshot animals have somewhat longer and more pointed snouts than normals. In side view, the upper lip droops and hangs down over the mandible; less of the rima ori is thus visible than in normals. This drooping of the upper lip is the sign by which newly born animals can be recognized as overshot.

On retracting the lips from the teeth, a very abnormal occlusion of the teeth is seen. In a normal animal, the lower incisors bite closely against the tubercula dentalia on the lingual surface of the upper incisors. The lower canines fit into the gap between the 3rd upper incisors and the upper canines. In overshot animals, there is no contact between the upper and lower incisors at all. The mandible is about 12–18 mm. too short in proportion to the upper jaw. The lower canines bite into the gap between the upper canines and the first premolars, instead of into the gap between the third incisors and the canines. This faulty occlusion is evidently the direct cause for the altered position of some of the teeth. The lower canines occlude with a region of the upper jaw which is wider than the normal region in front of the upper canines; the lower canines therefore point more outwards in lateral direction and at the same time are more curved backwards. The gap between upper canines and first upper premolars is wider than normal; this is apparently due to the lower canines, which in the abnormal animals come to lie between these teeth when the jaws are closed. On the other hand, the distance between the lower canines and the first lower premolars is rather shorter than in normals. The front parts of the os incisivum are bent downwards, and the upper incisors are pointing downwards and backwards; this is evidently due to the lack of counter-pressure normally exerted by the lower incisors. As a consequence, the gap between the third incisors and the canines is



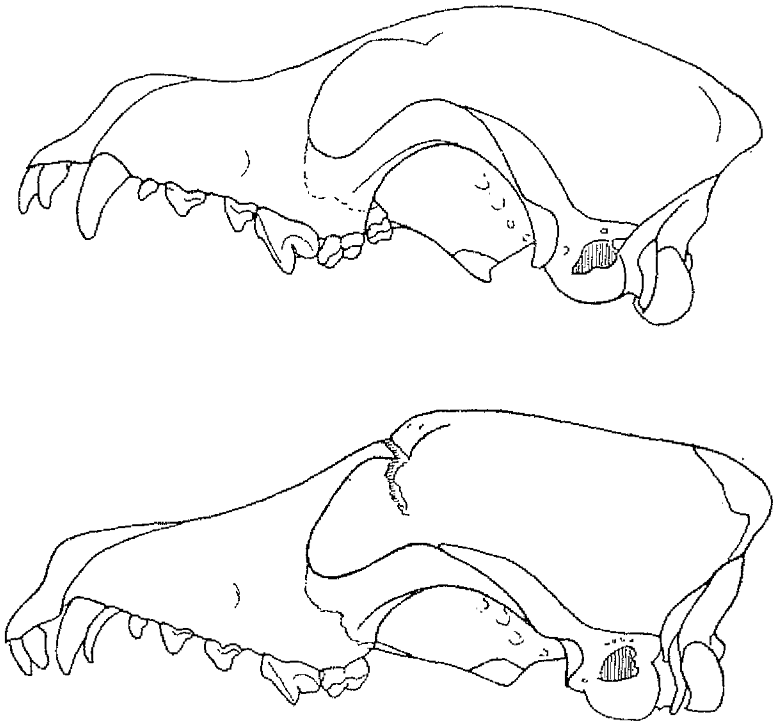
Text-fig. 1. Normal skull (bitch, above) and abnormal skull (dog, below). Note faulty occlusion of the front teeth, presence of persistent upper milk canine and absence of second upper molars in the abnormal animal. The flatter outline of the neurocranium in the abnormal dog is due to a somewhat stronger occipital crest and probably accidental. All drawings are approximately 0·58 natural size.

narrower than normal, and the anterior part of the hard palate is considerably more concave than in a normal animal.

In striking contrast to all these anomalies of occlusion in the front teeth, the occlusion in the region of the carnassials and molars is entirely normal, both in the living animals and in the skulls. This suggests that the anomaly of the overshot animals concerns the anterior parts of the jaws rather than the jaws as a whole.

We mention here some differences as regards the teeth between the normal and abnormal skull. Most of them seem to be "accidental":

(1) Presence of persistent milk canines. A persistent upper milk canine behind the permanent tooth is present on the left side of the abnormal dog, and an empty socket on the right side indicates that a similar tooth was originally present there. A similar condition existed

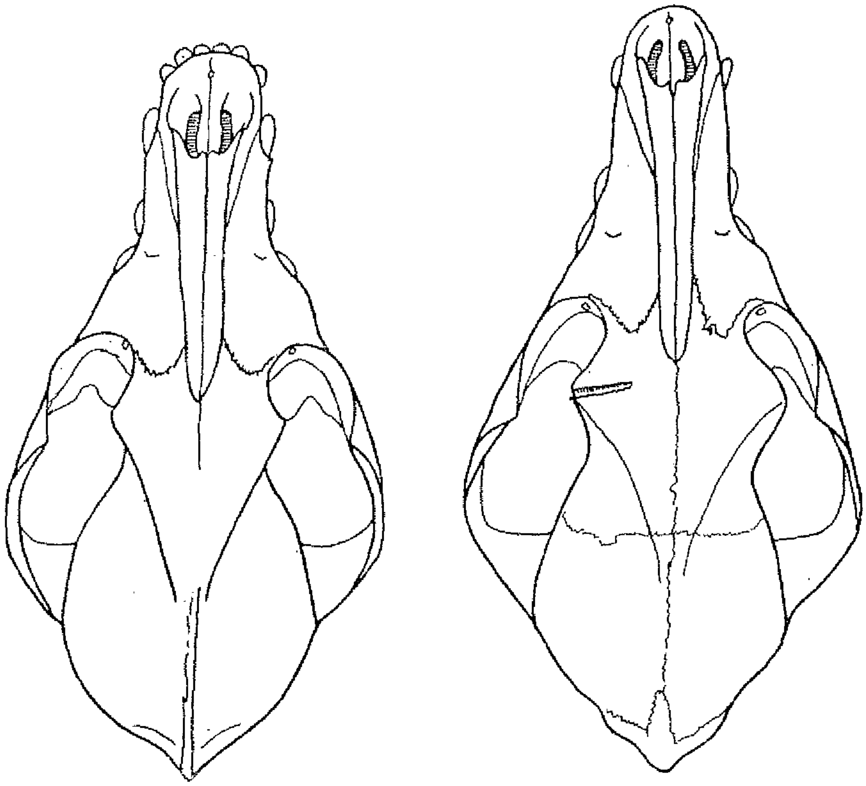


Text-fig. 2. Normal skull (bitch, above) and abnormal skull (dog, below). Note the bending down of the os incisivum, and the difference in shape of the pterygoid process. The latter difference may or may not be accidental.

for a short time in the abnormal bitch, but the milk canines were eventually shed. Persistence of upper milk canines is not rare in dachshunds in general. Their occurrence in the abnormal skull is thus probably accidental.

(2) Bilateral lack of second upper molars in the abnormal skull. Both the living normal and abnormal have the upper second molars, as shown by X-ray photographs. These teeth are not constant in dogs in general. Of eighty-five dog skulls in the collection of the Department of Zoology, University College, London, the tooth was absent bilaterally in two

individuals and hemilaterally in another animal. It may be mentioned that in the same collection, a third upper molar was present bilaterally in two and hemilaterally in one individual; bilateral occurrence of five instead of four premolars in the maxilla was found in one animal.

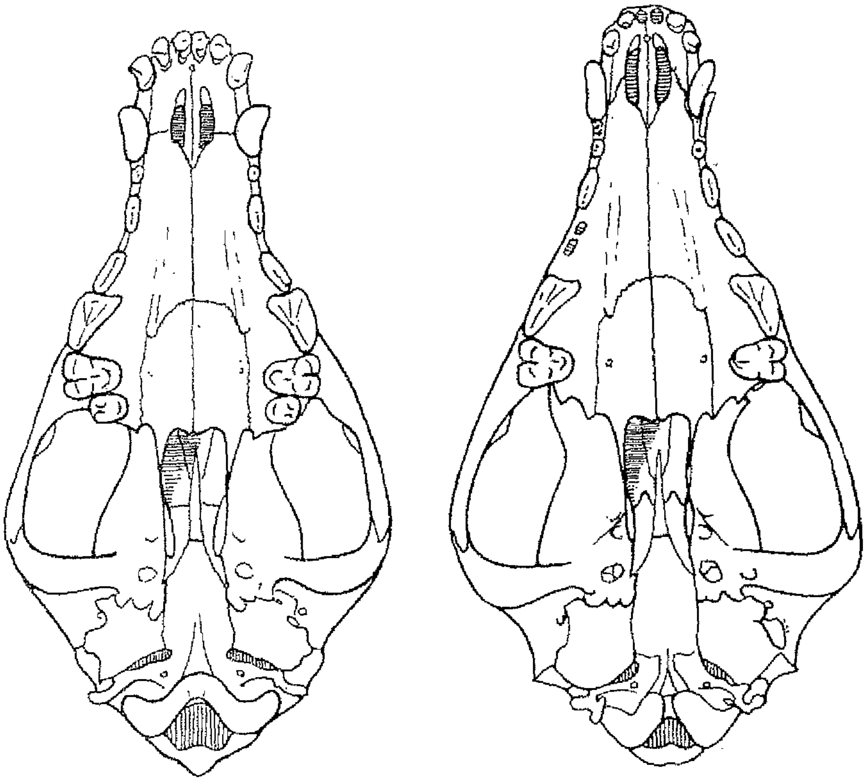


Text-fig. 3. Top views of normal skull (bitch, left) and abnormal skull (dog, right). Note the increased length of the nasal bones in the abnormal individual, and the fact that several cranial sutures are still visible which have disappeared in the normal animal. The abnormal skull is wider in the region of the frontal bones; this difference is accidental and may be a sex difference.

(3) Normal dogs have an accessory cusp at the distal ridge of the second, third and fourth premolars in the mandible, the cusp being biggest in the fourth and smallest in the second premolar; similar but smaller cusps occur on the second and third premolar in the maxilla. In the abnormal skull these cusps are very much reduced in size; in the lower jaw, the cusp of the fourth premolar is much smaller than normal, and those of the second and third premolar are absent altogether. The

cusps in the maxillary teeth are virtually absent. The condition resembles very much the normal appearance of these teeth as found in the fox.

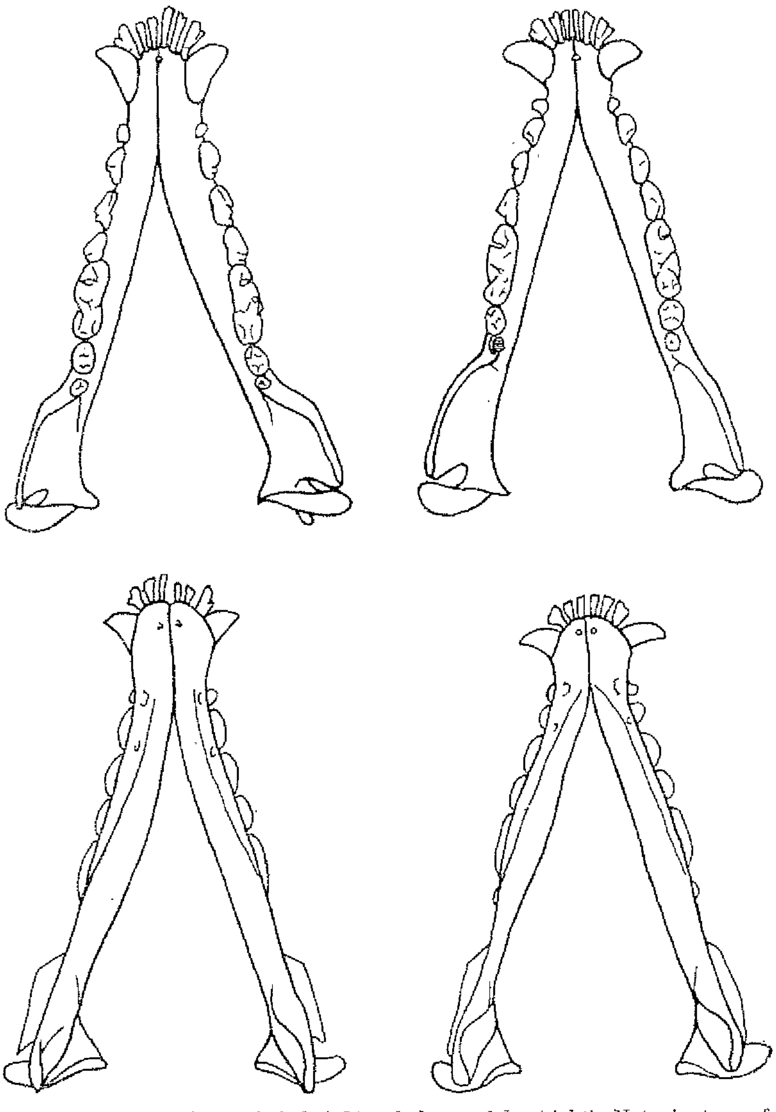
The premolars of the living abnormal bitch and the normal dog are completely normal. There is thus little doubt that the abnormality of these teeth as found in the skull of the abnormal dog is unconnected with



Text-fig. 4. Skull basis view of normal bitch (left) and abnormal dog (right). Note particularly the difference in the direction of the sutures between the ossa incisiva and the maxillae, proving lengthening of the maxilla in its anterior parts. The foramina palatina anteriora are bigger in the abnormal animal (typical difference). The distance between third premolar and carnassial is greater in the abnormal animal (? accidental difference). Several minor differences in the region of the pterygoid and the otic region are of doubtful significance.

the jaw anomaly. On the other hand, the perfectly regular appearance of the cusp reduction strongly suggests that it may be genetic in origin. The appearance of these cusps in dogs in general is somewhat variable. In the skull collection referred to above, no individual with a similarly complete reduction of these cusps has been found.

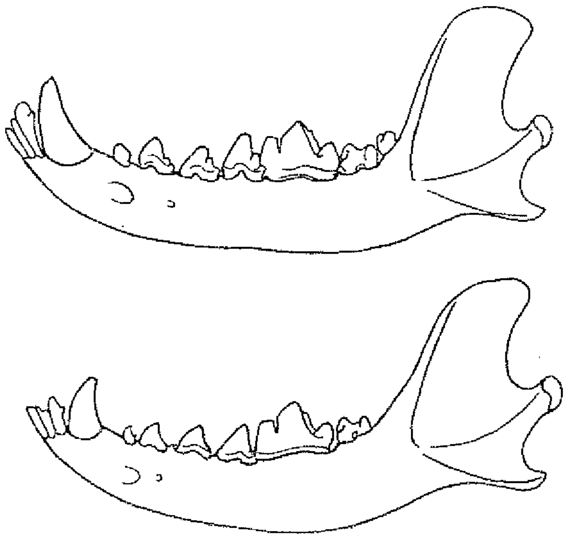
All the teeth are considerably smaller in the abnormal skull than in the normal one. A similar difference is found between the teeth of the two living animals. This difference may be typical. But tooth size is very



Text-fig. 5. Mandible of normal bitch (left) and abnormal dog (right). Note shortness of the symphysis in the abnormal animal.

variable in dogs in general and in dachshunds in particular. We therefore cannot exclude the possibility that the resemblance between the two normals on the one hand and the two abnormals on the other hand, may be accidental.

The following description is mainly based on the skulls; the main features are also recognizable in the radiographs. In the abnormal animal, the snout is somewhat elongated. This is particularly noticeable in the nasals and to a somewhat lesser extent in the maxilla. The os incisivum does not appear to differ in size from the normal, apart from the fact that its anterior part with the incisors is bent downwards. The dorso-ventral diameter of the nose, that is to say, the distance between its roof



Text-fig. 6. Mandible of normal bitch (above) and abnormal dog (below), seen normal to the plane of the ramus mandibulae. Note closeness of the two foramina mentalia in the abnormal individual and reduction of the accessory cusps in the second, third and fourth premolar.

and the hard palate, is reduced. The foramina palatina anteriora are considerably longer anteriorly; this is also visible in the radiographs in top view. Similarly large foramina did not occur in the skull collection mentioned above.

The mandible is shorter than normal. The shortening seems to affect the anterior parts only. The symphysis is much shorter, and the two foramina mentalia on each side of the mandible are closer together. From the molar region backwards, no difference is noticeable between the normal and abnormal mandible.

It therefore appears that the faulty occlusion of the front teeth is caused both by a shortening of the mandible and a lengthening of the upper jaw, at least in fully grown animals. An investigation of the radiographs suggests that the mandible is primarily affected, and that the alterations in the upper jaw are of a secondary nature. In the earliest radiograph (47 days, Pl. IV), there is no appreciable difference in snout length, but the anterior parts of the mandible are already clearly shortened; the abnormal occlusion is precisely the same in the deciduous dentition as it is later found in the permanent teeth. Even at the age of 152 days, the lengthening of the upper jaw is not yet so marked as in the skull, and the bending down of the os incisivum has not yet so far developed. On the other hand, the reduced dorso-ventral diameter of the nose is already clearly noticeable at that age.

The following explanation seems plausible, though it is far from being proven. The mandible is primarily affected. This leads to an occlusion of the lower canines behind instead of in front of the upper canines. It seems that the lower canines exert pressure against the upper canines from behind and thus secondarily cause an elongation of the anterior parts of the upper jaw. The bending down of the os incisivum is due to the lack of counter-pressure, as mentioned above. Perhaps there is some relation between the reduced height of the nose and this bending down. Similarly, there is presumably some connexion between the abnormal strains and stresses caused by the abnormal occlusion and the increased size of the foramina palatina anteriora.

The neurocranium seems to be entirely unaffected by the jaw anomaly. It will be noticed that the width of the frontals, where they contain the frontal sinuses, is considerably greater in the abnormal skull. In the living animals, the reverse seems to hold, to judge from palpation (the region is not clearly visible in radiographs). The difference is thus "accidental"; as it differentiates the two dogs from the two bitches, it may be a secondary sexual character. Minor differences in the pterygoid processes, in the shape of the foramen occipitale magnum and the atlases may or may not be accidental; these structures are not clearly visible in the radiographs, and no comparison is thus possible. The same applies to a difference in the obliteration of the cranial sutures. These are far more obliterated in the normal skull than in the abnormal one. No other bones of the skeleton have been available for examination.

The question arises as to the cause of the reduced growth of the anterior parts of the mandible. No definite answer can be given, but a tentative suggestion may be made. It was pointed out above that both abnormal animals have considerably smaller teeth than the normals. This is particularly striking in the case of the skull of the abnormal dog, which is bigger than that of the normal bitch. If this difference should turn out to be typical, it may be the root cause of the anomaly. It seems that in man, at any rate, the size of the maxilla is less dependent on the size of the teeth than the mandible. It is often found that the maxillary teeth stand in loose formation with gaps in between the teeth; in such cases the maxilla is evidently bigger than is required by the size of the teeth. No such loose formation is found in human mandibles, where the teeth always stand close together. If a corresponding relation holds for the dog, a general reduction of tooth size might lead to a shortening of the mandible without affecting the upper jaw.

Summary

A new autosomal recessive gene is described in long-haired dachshunds which causes a faulty occlusion of the front teeth. It appears that the primary effect of the gene results in a shortening of the anterior parts of the mandible. The faulty occlusion thus produced apparently leads to a lengthening of the upper jaw.

Acknowledgements

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EXPLANATION OF PLATE IV

X-ray photographs of normal dog (above) and abnormal bitch (below), litter mates, 47 days old. In both radiographs, parts of a human hand holding the animal's head are visible.

Reference

Haldane, J. B. S. (1938). "The estimation of the frequencies of recessive conditions in man." *Ann. Eugen., Lond.*, 8, 255–62.