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## Generalized Congenital Hypotrichosis in a Female Rottweiler

PETER J. IHRKE,\*† RALF S. MUELLER‡§ and ANTHONY A. STANNARD‡

\*Department of Medicine, †Veterinary Medical Teaching Hospital, School of Veterinary Medicine, University of California, Davis, CA 95616, U.S.A.

(Received 16 June 1993; accepted 23 August 1993)

**Abstract**—Generalized congenital hypotrichosis is reported in a female Rottweiler. This is only the second report of this genodermatosis in a female dog. The dog was predominately hairless except for bilaterally symmetric tufts of hair on the head, dorsal midline and umbilical region. Most skin biopsy sections revealed a total lack of epidermal appendages representing a complete failure of follicular development. Occasionally, small rudimentary follicular structures were subtended by melanophages and vertically oriented collagen suggesting the possibility of previously more extensive development followed by subsequent involution. According to a new classification scheme for ectodermal dysplasia proposed by Dr Carol Foil, this dog would be classified as 1–4 defect, cogenital hypotrichosis.

**Key Words:** Generalized congenital hypotrichosis; Ectodermal defect; Ectodermal dysplasia; Genodermatosis; Rottweiler.

### INTRODUCTION

Congenital hypotrichosis is a rare subgroup of genodermatoses, within the larger category of ectodermal dysplasia, characterized by varying degrees of alopecia present from birth due to a diminished number of adnexal structures. Lack of hair may be the only abnormality noted or may be seen concomitantly with other ectodermal defects such as abnormal dentition and decreased tear production (1–6). Congenital hypotrichosis has been previously documented convincingly in the whippet (1), cocker spaniel (2), Belgian shepherd dog (3), lhasa apso (5), miniature poodle (7), basset hound (8), beagle (9), labrador retriever (9), bichon frise (10) and the Yorkshire terrier (11). All previous reports of this syndrome, with the exception of a female labrador retriever reported by Kunkle (9) have been in male dogs implying sex-linked inheritance. Congenital lack of hair in a female Rottweiler is described in this report.

A 5-week-old, female Rottweiler was examined because of generalized absence of hair since birth.

†Correspondence and requests for reprints to Dr P. Ihrke at the above address.

‡Dr Mueller's current address is: Animal Skin and Allergy Clinic, 547 Dandenong Rd, Armadale, Victoria 3143, Australia.

The affected puppy initially was the same size as its siblings, but subsequently showed a slower growth rate. At presentation, the dog was predominately hairless (Fig. 1). Over 95 per cent of the body surface was affected. Tufts of short, thin, wirey hair were present only on the forehead, back of the neck, dorsal midline and around the umbilicus in a partially bilaterally symmetric pattern (Fig. 2). These hairs appeared grossly normal when viewed with a light microscope. Isolated, fine vellous hairs could be identified with a hand lens. Facial tactile sensory hairs were present but were sparse, short, wirey and of variable diameter. Eyelashes were not present. All nails were normal (Fig. 3). The area of exposed skin were variably grey, dry, and mildly scaly. A greasy odor was noted. All of the premolars were present, however, the canine teeth were not yet evident. Ocular abnormalities were not noted and the results of a Schirmer's tear test were normal. Other abnormalities were not noted on physical examination.

Blood was taken for a complete blood count, serum chemistry profile, and baseline serum thyroxine and triiodothyronine analyses. Multiple skin biopsy specimens were obtained, fixed in formalin, processed routinely, and stained with hematoxylin and eosin (H&E). Results of the complete blood count, serum chemistry profile values, and baseline serum thyroxine and triiodothyronine analyses were all within normal limits.

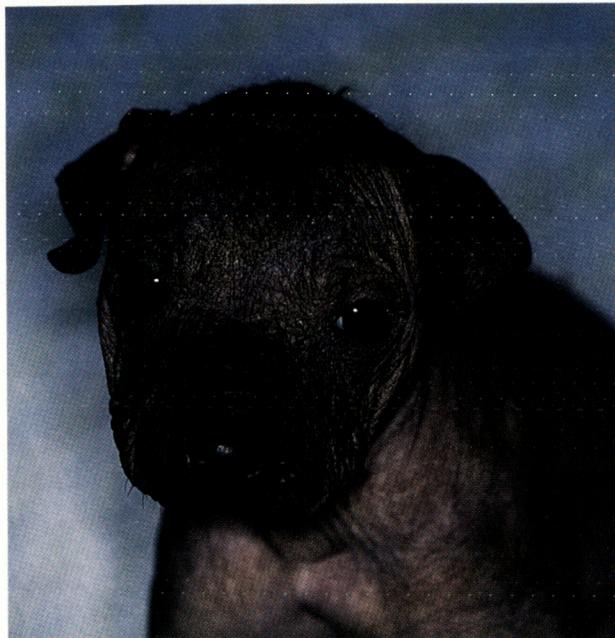


Figure 1. Congenital hypotrichosis in a Rottweiler at 5 weeks of age. Note sparse hair in the temporal region.



Figure 2. The ventrum is almost totally alopecic except for the tuft of hair surrounding the umbilicus.

Most skin biopsy sections revealed a total lack of epidermal appendages. Occasionally, small rudimentary follicular structures were observed. These varied from focal accumulations of small basophilic cells "budding" from the basal cell layer of the epidermis (Fig. 4) to recognizable follicular structures (Fig. 5). The former were thought to be analogous to the primary epithelial germ (hair germ) in the developing fetus. Neither sebaceous glands nor sweat glands were observed in any of the sections. The histopathological changes noted above in this dog were similar to those previously reported by Selmanowitz (7), and represent a more extensive ectodermal defect of the skin than is usually seen with congenital hypotrichosis.



Figure 3. Distal extremity is totally alopecic but nails are unaffected.

Congenital hypotrichosis has been assumed to be a partial to complete failure of follicular development. Most of our microscopic findings would support that view. However, the observation of melanophages and vertically oriented collagen below some of the rudimentary follicular structures would suggest the possibility of previously more extensive development followed by subsequent involution (Fig. 6). Recently, Cotsarelis has hypothesized that a failure of communication between the follicular stem cells located in the bulge area of the pilosebaceous unit and the subjacent dermal papilla may explain initial hair growth followed by follicular involution and tardive hypotrichosis (12). The possibility of such a phenomenon is supported by examples of tardive onset hypotrichosis reported in a variety of species. In these cases, the animal is born with varying degrees of haircoat that is subsequently lost during the first few months of life (13).

Recently, Foil has proposed a new and useful provisional classification scheme for ectodermal dysplasia (Table 1) (11). According to this classification scheme, the combined clinical and histopathologic features exhibited by this Rottweiller would lead to classification as a 1-4 defect congenital hypotrichosis since adnexal as well as hair follicle and hair shaft defects were noted.

Periodic reexaminations were performed during the 8 months following initial presentation. The degree of alopecia remained static (Figs 7-9). Gradual increases in hyperpigmentation and scaling were noted and the greasy odor became more pronounced. All nails remained normal and the adult dentition, although delayed, was normal. Adult size was within the breed standard for the Rottweiler giving the impression of a giant Chinese crested dog.

Five littermates of the affected bitch (two other females and three males), had normal haircoats. The pedigrees of the sire and dam were reviewed

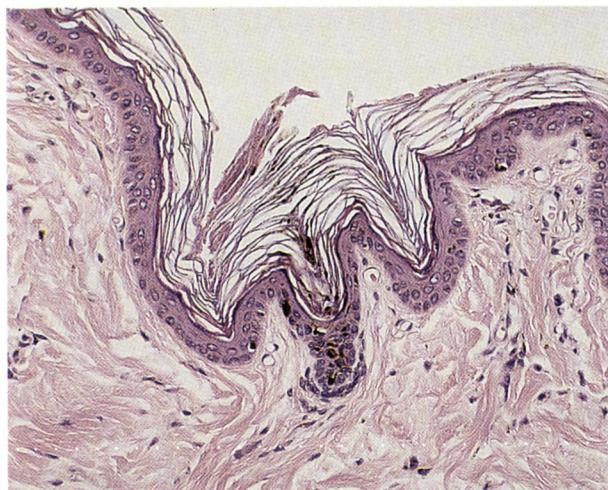


Figure 4. Focal accumulation of small basophilic cells "budding" from the basal cell layer of the epidermis (H&E  $\times 690$ ).

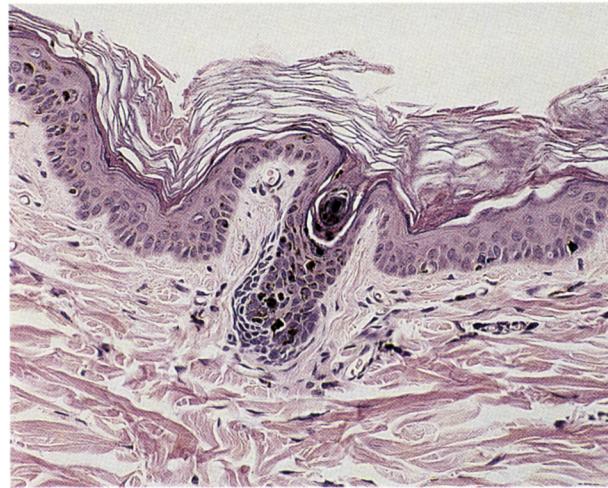


Figure 5. A solitary, rudimentary hair follicle. Note the prominent accumulation of melanin in the ostia in the absence of recognizable hair (H&E  $\times 690$ ).

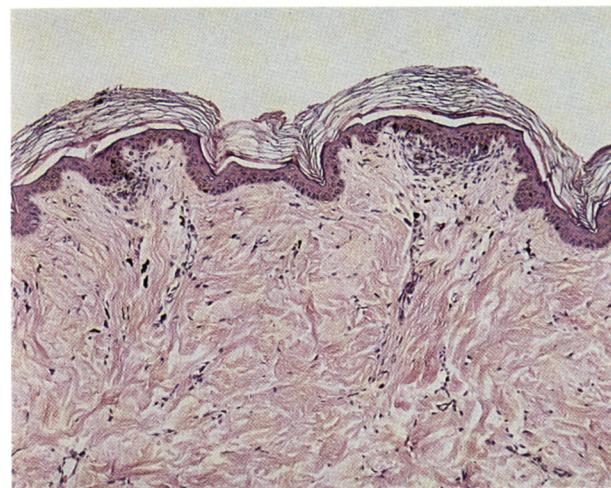


Figure 6. Vertically oriented collagen fibers and melanophagelike accumulations underlying "buds" similar to the one illustrated in Fig. 5 (H&E  $\times 345$ ).



Figure 7. Congenital hypotrichosis in a Rottweiler at 5 months of age. Note the increased hyperpigmentation.



Figure 8. Closer view of the face at 5 months of age. Sparse hair is present on the dorsal head and sensory hairs are evident.

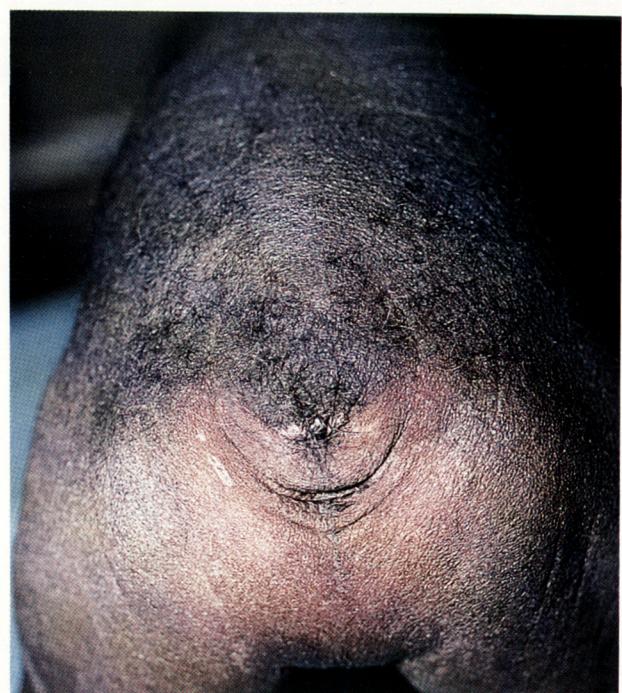


Figure 9. Closer view of the perianal region at 5 months of age. Scaling and hyperpigmentation have increased.

for four generations and duplication of ancestors was not seen. The male previously had sired normal litters and a prior breeding of the bitch to a different sire had yielded eleven normally-coated puppies.

The characteristic clinical feature of congenital hypotrichosis is a lack of hair in well delineated areas. The head, ears and ventrum are affected most commonly (1, 3, 7, 9). Remaining hair, when present, usually is bilaterally symmetric and is most commonly localized to the dorsum of the head, distal limbs and tail, umbilical zone and areas surrounding mucocutaneous junctions (5, 13). Selmanowitz has hypothesized that skin appendages in these haired areas may be under different morphogenetic influences, perhaps of neural crest origin, than in other body regions (13). These different morphogenetic influences could lead to the uniformly distinctive bilaterally symmetric patterns seen in the remaining haircoat of dogs with more generalized hypotrichosis.

Both localized and generalized congenital hypotrichosis are likely the result of a similar genetic phenomena. Near total hairlessness is thought to be a recessive trait (4). Sex-linked inheritance usually has been implicated due to the preponderance of male dogs reported with congenital hypotrichosis. Spontaneous mutations also probably account for the hairless breed of dogs (5). Presumably, the initial hairless dogs used to develop the hairless breeds exhibited generalized congenital hypotrichosis as a spontaneous mutation. Since neither localized nor generalized congenital hypotrichosis have been reported previously in the Rottweiler, spontaneous mutation is a likely explanation.

The distinctive pattern of hairlessness seen in this dog mirrors the pattern seen in canine alopecic breeds such as the Chinese crested dog, Mexican hairless dog, Abyssinian dog, Turkish naked dog and the African sand dog. Interestingly, O'Neill (Foil) has published a photograph of a Lhasa Apso with generalized congenital hypotrichosis, seen by Kunkle, with a pattern of hairlessness that is virtually identical to that of the Rottweiler in this report and many of the established alopecic breeds (5). This adds credence to the theory espoused by Selmanowitz that skin appendages in different body areas may be under different morphogenetic influences.

The diagnosis of congenital hypotrichosis requires that the affected animal have a history of absence of hair since birth or shortly after birth and that the alopecia must not be progressive. In confirmatory clinical history denoting these criteria is lacking, congenital hypotrichosis must be differentiated from other ectodermal dysplasias such as canine follicular dysplasia and skin diseases characterized by follicular atrophy such as the endocrinopathies. Histopathology would be required to confirm the diagnosis.

TABLE 1. Provisional classification of ectodermal dysplasia in dogs and cats<sup>†</sup>

1 = hair defect
1a = color-linked
1* = associated with abnormal pigment granules (Maltese dilution)
2 = tooth defect
3 = defect in claws (no heritable defects yet described)
4 = adnexal gland defect
5 = other ectodermal structural defects (ocular, olfactory, auditory, nervous system)
<b>1-2-4 Defects</b>
Chinese Crested Dog, Mexican Hairless Dog
<b>Congenital Sex-linked ectodermal defect</b> —Miniature Poodle
<b>Congenital ectodermal defect</b> —Lhasa Apso, Yorkshire Terrier
<b>1-4 Defects</b>
<b>Congenital hypotrichosis</b> —Cocker Spaniel, Whippet, Belgian Shepherd, French Bulldog, Rottweiler
<b>1 Defects</b>
<b>Congenital hypotrichosis</b> —Beagle
<b>Canine Follicular Dysplasia</b> —Siberian Husky, English Springer Spaniel, Airedale Terrier, Alaskan Malamute, Irish Water Spaniel, Portuguese Water Dog, Curly-Coated Retriever, German Short-Haired and Wire-Haired Pointers, Chesapeake Bay Retriever
<b>Flank Alopecia</b> —Boxer, English Bulldog, French Bulldog, Miniature Schnauzer
<b>Acquired Pattern Alopecia</b> —Miniature Dachshund, Boston Terrier, Chihuahua, Whippet, Manchester Terrier, Italian Greyhound
<b>Canine alopecia universalis</b> —Beagle, American Hairless Terrier (Rat Terrier)
<b>Feline alopecia universalis</b> —Siamese
<b>Feline tardive hypotrichosis</b> —Siamese
<b>Feline lethal(?) pili torti</b> —domestic short-haired cat
<b>Feline recessive alopecia universalis</b> —Sphynx cat, Cornish and Devon rex cat, American wire-haired cat
<b>1a Defects</b>
<b>Congenital hypotrichosis</b> —Yorkshire Terrier
<b>1a* Defects</b>
<b>Congenital (sex-linked?) hypotrichosis</b> —Basset Hound, silver Miniature Poodle
<b>Color dilution alopecia</b> —Doberman Pinscher, Dachshund, Great Dane, Whippet, Standard Poodle, Chow Chow, Irish Setter, Italian Greyhound, Boston Terrier, Chihuahua, Yorkshire Terrier, Saluki, Miniature Pinscher, mixed-breed dogs (some may have 1a*-4 defects)
<b>Black-hair follicular dysplasia</b> —mixed-breed dogs, Bearded Collies, Basset Hounds, Papillons, Schipperke, Beagle, Saluki, Cocker Spaniel, Gordon Setter
<b>1-4 Defects</b>
<b>Feline lethal recessive hypotrichosis</b> —Birman
<b>1-2-4-5 Defects</b>
<b>Congenital hypotrichosis</b> —Labrador Retriever
<b>1-4-5 Defects</b>
<b>Congenital alopecia</b> —Bichon Frise

<sup>†</sup>Table adapted from Foil, C. In: Hoskins, J. D. ed. *Veterinary Pediatrics*. Philadelphia: W. B. Saunders, 1993: 366. Reproduced by permission of W. B. Saunders Co.

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**Résumé**—Une hypotrichose congénitale généralisée chez une chienne Rottweiler est rapportée. Il s'agit du second cas de cette génodermatose décrit chez une chienne. Le chien était nu exception faite de touffe de poils sur les flancs et la tête, la ligne du dos et la région ombilicale. La plupart des biopsies ont révélé une absence totale d'annexe et de développement folliculaire. Parfois quelques structures folliculaires rudimentaires étaient subtentés par la présence de mélanophages et une orientation verticale des fibres de collagène. En suivant la nouvelle classification des dysplasies ectodermiques proposée par le Dr Carol Foil, cet animal a été classé en hypotrichose congénitale de défaut 1-4. [Ihrke, P. J., Mueller, R. S., Stannard, A. A. Generalized congenital hypotrichosis in a female Rottweiler (Hypotrichose congénitale généralisée chez une chiènne Rottweiler). *Veterinary Dermatology* 1993; 4: 65-69].

**Resumen**—En éste artículo se describe un caso de hipotricosis congénita de tipo generalizado en una hembra de la raza Rottweiler. Éste es el segundo caso de genodermatosi reportado en una perra. Éste animal padecía de una pérdida de pelo general exceptuando algunas mechas aisladas en la cabeza, línea media dorsal y región umbilical, de una manera bilateral y simétrica. La mayoría de las biopsias cutáneas revelaron una falta total de apéndices cutáneos, lo cual representaba una fallo completo en el desarrollo del folículo. De manera espóradica, se observaron estructuras foliculares rudimentarias invadidas de melanófagos y con fibras de colágeno orientadas verticalmente sugiriendo la posibilidad de desarrollo previo mas extensivo seguido de posterior involución. De acuerdo con la nueva clasificación de displasia ectodérmica propuesta por Foil, éste animal sería clasificado con un defecto de 1-4 de hipotricosis congénital. [Ihrke, P. J., Mueller, R. S., Stannard, A. A. Generalized congenital hypotrichosis in a female Rottweiler (Hipotricosis congénital de tipo generalizado en una hembra Rottweiler). *Veterinary Dermatology* 1993; 4: 65-69].