

Acral mutilation and analgesia in 13 French spaniels

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(Received 14 January 2004; accepted 9 January 2005)

Abstract Acral mutilation and analgesia (AMA) is reported in 13 French spaniels in Canada. This newly recognized disorder shares striking similarities in clinical features and biopsy findings to the other acral mutilation syndromes or hereditary sensory neuropathies reported in German short-haired pointer dogs, English pointer dogs and English springer spaniels. Clinical signs are first noted between 3.5 and 12 months of age. Affected dogs lick, bite and severely self-mutilate their distal extremities resulting in ulcers with secondary bacterial infection. Auto-amputation of claws, digits and footpads occurs in severe cases. Single or multiple feet can be affected. Affected dogs walked on their severely mutilated feet without evidence of pain, lameness, or ataxia. The majority of the dogs were euthanized within days to months of diagnosis.

INTRODUCTION

Acral mutilation and analgesia (AMA) is a rare canine hereditary sensory neuropathy reported in over 100 German short-haired pointers from Europe, several English pointers from North America and English springer spaniels in Australia.^{1–10} The syndrome results in progressive mutilation of the distal extremities. Age of onset varies from 2 to 12 months with an average of 4 months. Clinical signs consist of sudden intense licking, biting and severe self-mutilation of the feet. If affected dogs are not restrained, auto-amputation of claws, digits and footpads usually results. One or several feet can be affected. Remarkably, affected dogs walk and even run without evidence of lameness or discomfort. Euthanasia is usually requested by owners. This disorder appears to be inherited in an autosomal recessive mode of transmission.

The authors have seen a disease with marked clinicopathological similarities in several French spaniels since 1994. The purpose of this study is to describe this syndrome in French spaniels.

MATERIALS AND METHODS

Data were obtained from the owners and breeders of affected dogs in the Province of Quebec, Canada. Dogs were selected based on compatible history and clinical features. Ten of the 13 dogs reported here were examined

by the Dermatology Service of the Veterinary Teaching Hospital (VTH) of the University of Montreal (UM). Clinical data were collected from all dogs. Pedigrees were obtained for each dog, data were collected referable to affected and unaffected relatives, and an ancestral tree was established.

Complete necropsy was performed in five dogs (dogs 1, 4, 8, 9 and 12). In addition, biopsies of mutilated feet (dogs 1 and 4) were performed prior to euthanasia. All tissues were routinely processed for histopathology and stained with haematoxylin–phloxin–safran; selected sections were stained with Luxol fast blue–cresyl echt violet, Bielschowsky, Gram and periodic acid Schiff.

Cytology, bacterial cultures and sensitivity, and fungal cultures were obtained from self-mutilated ulcers from six, three and two dogs, respectively. Neurological examination was performed in nine dogs and radiographs of mutilated feet were obtained in five dogs.

RESULTS

Familial history and clinical findings

Thirteen French spaniels (six females and seven males) from five different litters were examined. Pedigree analyses revealed that all 13 dogs shared common ancestors (Fig. 1).

None of the sires or dams of affected dogs were clinically affected. Sire A produced four of five affected litters: litter 1 with bitch A, and litters 3, 4 and 5 with bitch B (Table 1). Sire A had produced another three litters with two other females with no documented cases of AMA among the offspring. Bitch B had another litter with a different male with no known affected offspring. Sire B produced litter 2 with bitch C (Table 1). However,

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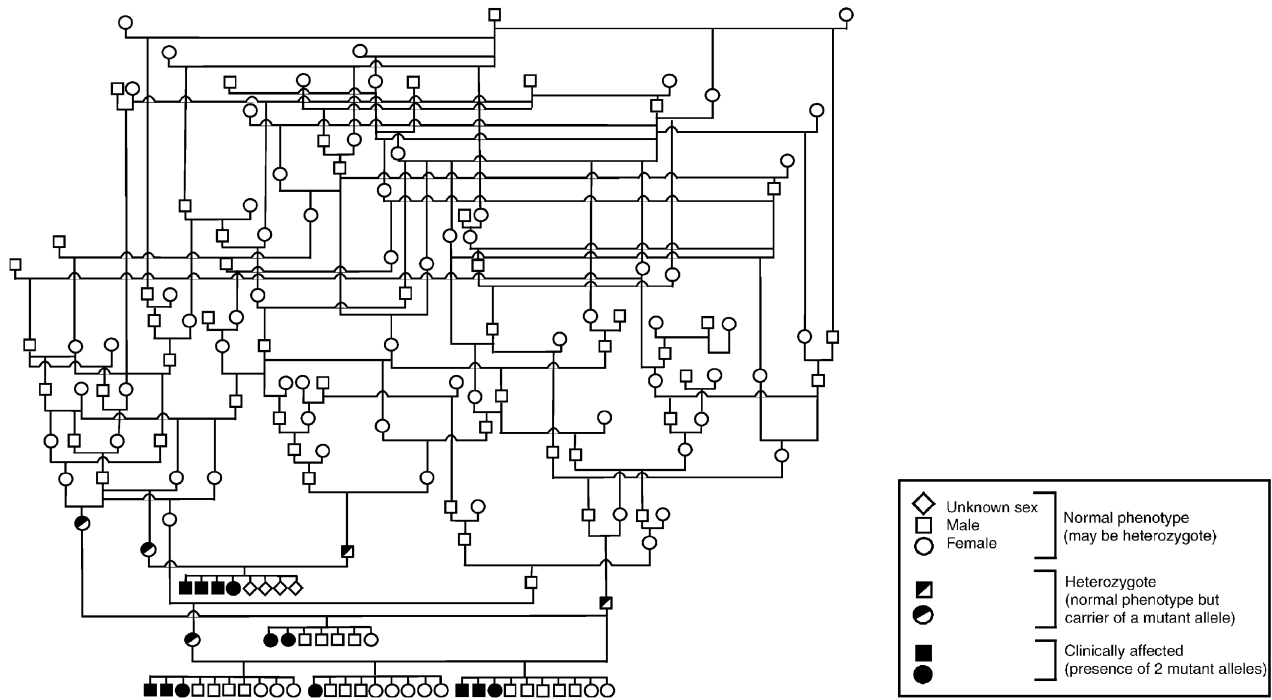


Figure 1. Ancestor tree of 13 French spaniels with acral mutilation and analgesia due to hereditary sensory neuropathy.

Table 1. Data and outcome of the 13 French spaniels with acral mutilation and analgesia due to presumptive hereditary sensory neuropathy

| Case number | Sex | Age at onset (months) | Age at diagnosis (months) | Number of affected feet at time of diagnosis | Outcome |
|-------------|-----|-----------------------|---------------------------|--|---|
| Litter 1 | | | | | |
| 1 | F | ≈ 9 | 14 | 3 (LH, RH, LF); 4 digits | Euthanized at 14 months |
| 2 | F | 10.5 | 13 | 2 (RF + RH); 2 digits and 1 footpad | Lost follow up |
| Litter 2 | | | | | |
| 3 | F | ≈ 3.5 | 11 | 2 (LH + RH); 1 digit, 2 footpads | Still alive at 8 years. No physical restraint and no problem since 3.5 years of age |
| 4 | M | 8 | 11 | 4; several digits and footpads | Euthanized at 11 months |
| 5 | M | ≈ 7 | 8 | 2 (RH + LH); 2 digits | Lost to follow up |
| 6 | M | ≈ 4 | 10 | 3 (RH, RF, LF); several digits and footpads | Euthanized at 11 months |
| Litter 3 | | | | | |
| 7 | M | 9 | 24 | 4; several digits and footpads | Still alive at 4.5 years. No physical restraint and no problem since 3 years of age |
| 8 | M | 3.5 | 4 | 1 (RF); 1 digit, 1 footpad | Euthanized at 4 months |
| 9 | F | 5 | 5.5 | 3 (RF, LH, RH); several digits and footpads | Euthanized at 5.5 months |
| Litter 4 | | | | | |
| 10 | F | 5 | 8.5 | 2 (LH, RH); 1 digit, 2 footpads | Euthanized at 8.5 month |
| Litter 5 | | | | | |
| 11 | F | 6 | 7 | 1 (LF); 1 footpad; 2 more feet (RF, RH) were affected 2 months later; several digit and footpads | Still alive at 2.5 years; No physical restraint and no problem since 2 years of age |
| 12 | M | ≈ 7 | 11 | 2 (RH + RF); 3 footpads and 2 digits | Euthanized at 11.5 months |
| 13 | M | 12 | 12 | 2 (LF,RH); 4 digits | Still alive at 2.5 years; still needs to wear E-collar most of the time |

F, female; M, male; LH, left hind foot; RH, right hind foot; LF, left fore foot; RF, right fore foot; Same sire (Sire A) for litters 1, 3, 4 and 5; Same bitch (bitch B) for litters 3, 4 and 5.

no affected offspring were identified from several other litters he had produced with several other bitches.

Case 1. A 9-month-old, intact female French spaniel was examined for two severely mutilated hind feet. The dog constantly licked and bit the affected feet. Radiographs revealed osteomyelitis and bony proliferation with lysis of most of the third phalanx of the second digits of both affected feet. Cytological examination from the ulcers revealed degenerated neutrophils and numerous phagocytized cocci. Bacterial culture and sensitivity revealed *Staphylococcus intermedius* sensitive

to various antibiotics including cephalexin. Fungal culture was negative. Treatment with oral cephalexin, at 30 mg/kg twice daily and daily wound dressing with silver sulfadiazine and bandages was initiated. The dog was transferred to the Surgery Service 2 days later for surgical amputation of affected digits. During hospitalization, it became obvious that the dog was insensitive to pain as discomfort was not elicited by wound care or bandaging and she walked normally on both severely affected extremities. The dog managed to destroy E-collars, remove bandages and skin sutures, create wound dehiscence and expose underlying bone.

Repeat cultures revealed the presumed contaminant, *Escherichia coli* also sensitive to cephalexin but no fungi.

The dog was eventually sent home with daily wound dressings and bandages with topical silver sulfadiazine, cephalexin to be administered orally, and a stronger E-collar made from a plastic bucket. Healing was protracted and the owners finally elected euthanasia 4 months later after new mutilation occurred on the right front foot.

Case 2. A 13-month-old, intact female French spaniel (sibling of dog 1) was referred with a 3-month history of right front foot mutilation that had started after boarding. Antibiotics and bandaging yielded minimal improvement. Several days prior to presentation, she had started chewing her right metatarsal footpad. Physical examination revealed swollen and ulcerated digits IV and V of the right front foot and a deep ulcer of the metatarsal footpad of the right hind foot. There was no evidence of osteomyelitis on radiographic examination. Cytological examination of the lesions revealed degenerate neutrophils, numerous phagocytized cocci and a few macrophages.

As for case 1, the dog walked normally on her mutilated feet without evidence of pain or discomfort. A presumptive diagnosis of AMA was made based on history and clinical findings. The owner was instructed to administer cephalexin orally for a minimum of 3 weeks, to change bandages daily and to fit an E-collar at all times in order to prevent licking and mutilation. The dog was lost to follow-up.

Cases 3–13. Information on cases 3–13 from litters 2–5, as well as additional information on dogs from litter 1, is summarized in Table 1. The striking clinical features shared by all 13 dogs were acral mutilation and apparent analgesia. Affected dogs began licking and biting digits and/or footpads of one to multiple feet at between 3.5 and 12 months of age. Rapidly progressing self-mutilating acral changes included swollen digits, paronychia, claw loss, footpad ulceration, digital amputation and occasionally fracture and osteomyelitis (Figs 2–5). Even in the more severe cases, the affected dogs walked and ran on mutilated feet without any



Figure 2. Case 9. Foot mutilation resulting in auto-amputation of the third phalanx of the fifth digit of the right fore foot.



Figure 3. Case 12. Note the ulcerated metatarsal footpad of the right hind limb and less severe lesion of a digital footpad.



Figure 4. Case 12. Severely wounded third and fifth digits of the right fore foot.



Figure 5. Case 12. Note the second phalanx exposed on the third digit of the right fore foot.

evidence of lameness or pain, and allowed wound care without evidence of pain or discomfort. All 13 dogs were otherwise healthy, with the exception of secondary bacterial infection of the wounded feet at some point for each dog. One dog (case 11) was apparently the smallest of the litter but she had a normal growth curve. All affected dogs that reached adulthood achieved normal adult bodyweight.

Neurological findings

Complete neurological examinations were performed in nine dogs; findings were similar. Pain response was not elicited from affected distal limbs by pricking or pinching, with the exception of two dogs (dogs 2 and 13) that appeared to have deep pain response to toe pinching. Typically, insensibility to painful stimuli was marked in the distal limbs and appeared to recede proximal to the carpus and tarsus. Remarkably, affected dogs walked normally on their mutilated feet with no evidence of pain, discomfort, lameness or ataxia.

Other sensory modalities, to the extent that they could be evaluated clinically, appeared intact. All affected dogs had normal proprioception, intact tendon reflexes, and normal muscle tone; they were able to support weight and had no evidence of weakness or ataxia. Nociceptive loss was not detected elsewhere on the body, with the exception of one dog that did not respond to a pinch test with haemostats on the pinnae and lips (case 4). The limitation of neurological clinical findings to nociception loss was evocative of acral mutilation in Pointers due to hereditary sensory neuropathy. Electrodiagnostic studies were not performed.

Cytological examinations and bacterial and fungal cultures from wounds

Ulcers from six dogs (dog 2, 3, 5, 6, 7 and 10) were examined cytologically on one or multiple occasions. Results yielded degenerate neutrophils, phagocytosed cocci and occasional macrophages, with the addition of rods on one occasion in dog 3.

Bacterial cultures (four from three dogs) revealed *S. intermedius* (dogs 1, 3 and 7) and *E. coli* (dog 1), all sensitive to expected antibiotics. Dog 3 had *Pseudomonas* sp. at 11 months, which was sensitive to enrofloxacin. Fungal culture from dogs 1 and 13 were negative.

Radiograph examination of mutilated feet

Radiograph examination revealed osteomyelitis of the second and third phalanx of the third digit of right hind paw in dog 6, osteomyelitis of the first and second phalanx and absence of the third phalanx of the fifth digit as well as swelling and osteomyelitis of the second phalanx of the second digit and swelling of the fourth digit of the right front paw of dog 8. Radiographs of the left hind foot of dog 3 at 3.5 months revealed four fractured metatarsi that had occurred 1.5 months prior to the onset of self-mutilation, and intertarsal subluxation at 6 months. Interestingly, the dog walked normally on the affected foot at all times.

Treatment and outcome

Topical treatment, bandages and systemic antibiotics had been administered intermittently to 11 of the 13 dogs. An E-collar and/or bandages were required in order to avoid further mutilation. Eleven dogs received various courses of antibiotics given orally (cephalexin, occasionally sulfamethoxazole-trimethoprim, enrofloxacin) for secondary bacterial infection. The lesions typically required 2–4 months to heal, if further

mutilation was prevented. Dogs 3, 4, 6, 7, 10 and 11 developed several new self-traumatic ulcers when left unattended. Some owners noticed that onset coincided with boarding or other potentially triggering factors (e.g. stress or nervousness, hard surface such as concrete, tendency to dig through kennel door with their insensitive feet). Unfortunately, despite close surveillance, almost constant use of mechanical restraint and efforts in limiting trauma to feet (soft surface, limited exercise), dogs almost always had recurrences. At best, dogs had only a few months without creating new lesions. Management of affected dogs was difficult, time-consuming, expensive and frustrating. Seven of the 13 dogs were euthanized at owner request. Four dogs were still alive 2–7 years after diagnosis, and two dogs were lost to follow up (Table 1). Typically, dogs that survived to adulthood continued to attempt to mutilate their feet whenever restraining devices were removed. However in two dogs, the tendency to mutilate appeared to attenuate over time. Indeed, at the time of writing, dogs 3 and 7 have been 4.5 and 1.5 years, respectively, without lesions or mechanical restraint (Table 1).

Pathological findings

Complete necropsy was performed on dogs 1, 4, 8, 9 and 12. The only gross lesions reported were the acral changes described above. The spinal cords and roots (with ganglia) were removed and fixed in 10% neutral-buffered formalin in all cases. Proximal and distal portions of peripheral nerves (dogs 4, 8 and 9), and cutaneous nerves from affected digits (dogs 8, 9 and 12) also were sampled. Sections of spinal cords, nerve roots, cervical, thoracic and lumbar dorsal root ganglia and peripheral nerves were processed routinely for histopathology, and stained with haematoxylin–phloxin–safran; selected sections were stained with Luxol fast blue–cresyl echt violet and Bielchowsky. A separate age- and weight-matched control dog was used for comparison of spinal dorsal root ganglia in all dogs. No changes were observed in any neurological tissue examined. The overall number of neurones in dorsal root ganglia appeared adequate. However, whole ganglia were not available for serial sectioning and neuronal numbers could not be objectively evaluated; thus, a mild neuronal loss may have been missed.

Skin biopsies performed on mutilated feet of dogs 1 and 4 (ante-mortem) and histopathological examination of affected digits of dogs 1, 4, 8, 9 and 12 (post mortem) revealed chronic ulcers consistent with self-mutilation. Neither deep bacterial or fungal infection nor neoplasia was found.

DISCUSSION

A syndrome characterized of acral mutilation characterized as ‘toe necrosis’ or ‘ulcero-mutilating acropathy’ was reported in German short-haired pointer dogs from Europe between 1951 and 1974.^{1–4} Analyses of pedigrees indicated a recessively inherited disorder.⁴ The syndrome

described in German short-haired pointer dogs appears similar, if not identical, to the hereditary sensory neuropathy described in English pointer dogs from North America in the early 1980s.⁵⁻⁷ The syndrome in English pointer dogs has been seen with increasing incidence and geographical distribution.¹¹ Limited pedigree data suggests an autosomal recessive mode of inheritance. Recently, acral mutilation was reported in five English springer spaniels from Australia.^{8,9} Simple segregation analysis also suggests an autosomal recessive mode of inheritance in this breed.

The disorder in French spaniels reported here shares striking similarities in clinical features and biopsy findings to the other acral mutilation syndromes or hereditary sensory neuropathies reported in German short-haired pointer dogs, English pointer dogs, and English springer spaniels.

Interestingly, the French spaniel as one of the oldest pointing breeds (dating from the 17th century), may have contributed to the creation of other pointer breeds.¹² In the early 1900s, pointer 'blood' was introduced to the English springer spaniels.⁹ Moreover, after World War II, French spaniels were a disappearing breed with fewer than 50 dogs recorded in Europe. Out-crossing with other pointing breeds was performed to regenerate the breed.

Most neuropathies cause impairment of both motor and sensory function.¹³ In the disorder reported here, only nociceptive function appears affected. Our inability to elicit pain in the distal limbs suggests a syndrome affecting the nociceptive component of the peripheral nervous system. The absence of weakness, the presence of intact patellar reflexes, normal muscle tone and the ability to support weight provide further clinical evidence that motor fibres were unaffected.¹⁴

In dogs, the only sensory modalities that can be adequately evaluated are proprioception and nociception.¹³ Nociceptors, which are found in the skin, viscera and other tissues, are pain receptors that signal tissue damage or stimuli intense enough to threaten tissue damage. They are associated with both small myelinated (A δ) and unmyelinated (C) axons.¹³ The established method for evaluating nociception is to apply a haemostat to a 'tent' of raised skin. If there is no response, deeper structures including the digits should be evaluated. In animal with generalized loss (analgesia) or diminution (hypalgesia) of pain, it is recommended that the pinnae and face also be evaluated.¹³ It remains that in veterinary medicine, the nociceptive function is difficult to quantitatively evaluate.

Sensory neuropathies have been reported in other dog breeds.¹³⁻²¹ Canine sensory neuropathies present with a variety of clinical and pathological features, presumably reflecting diverse aetiologies. Most acquired or hereditary neuropathies reported in dogs are characterized by gait anomaly but without acral automutilation.¹³⁻²¹

The nociceptive loss and acral mutilation found in hereditary sensory neuropathy of English pointer dogs have been associated in one dog with deficiency of

primary sensory neurones.⁵ Changes included small ganglia with reduced numbers of cell bodies, and degeneration of unmyelinated and myelinated fibres in dorsal roots and peripheral nerves. The only spinal cord changes occurred in the dorsolateral fasciculus where reduced fibre density appeared to correlate with the observed nociceptive defect.⁵ The neuronal degeneration, however, appeared quantitatively inadequate to account for the deficiency of sensory cell bodies. It was concluded that this mutilation acropathy was a manifestation of a sensory neuropathy in which the neuronal deficiency results from insufficient development and slowly progressive postnatal degeneration.⁵

The pathological basis for this acral mutilation syndrome in French spaniels was not established. Further studies are needed to characterize whether this syndrome is a sensory neuropathy, as expected. Individuals with sensory loss of pain often die in childhood because they fail to notice injuries and illnesses.²² Diminished pain perception can result in decreased protection from external or self-inflicted damage.²³ Paraesthesia (burning, aching or lancinating pain), caused by aberrant sensory input to the central nervous system could account for the self-mutilation by these spaniels. However, it is impossible to test this in dogs.

Most inherited disorders of dogs have an autosomal recessive inheritance pattern.²⁴ Because both male and female French spaniels were clinically affected and all sires and dams that produced affected dogs were not clinically affected, it seems likely that the disease reported here also may be an autosomal recessive disorder. In addition, the number of affected dogs (29.5%, 13/44 dogs) was not significantly different from the expected incidence (25%) of autosomal recessive disorders.

The first French spaniels were imported into Canada in the 1970s, and the breed was granted Canadian Kennel Club recognition in the 1980s. In the early 1990s, two dogs (sire A and B) were imported from France. Our data suggests that these two males may have introduced this syndrome into Canada. French spaniels with syndromes resembling acral mutilation have been seen sporadically in France. One of the authors (MP) was recently consulted by veterinarians from France concerning two well-documented unrelated cases.

As cure is unlikely for acral mutilation and analgesia, the efforts of breeders, veterinarians and researchers must be directed toward prevention. In the absence of a carrier test, the recommendation to prevent recurrence of a recessive disorder is to remove affected animals and their parents from the breeding programme. The breeding of siblings should also be discouraged.

CONCLUSION

Acral mutilation and analgesia is a newly recognized disorder in French spaniels. Similar syndromes with markedly similar features in other related breeds suggest a sensory neuropathy. Preliminary genetic analysis suggests that transmission may be autosomal recessive.

The clinicopathological findings, the early age of onset and disease progression in affected French spaniels is very similar to hereditary sensory neuropathy reported in German short-haired pointers, English pointers and English springer spaniels. Acral mutilation syndromes are difficult to manage clinically and the majority of affected dogs are euthanized. Further studies are needed in order to more fully characterize the clinicopathological findings, aetiology, and likely genetic aspects of this disease.

ACKNOWLEDGEMENTS

The authors would like to thank Drs Claude Favrot, Peter Ihrke, Joan Parent, Jean-Martin Lapointe, Igor Mikaelian, Marie-Antoinette Perronne, Brigitte Siliart and David Silversides for their kind assistance.

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Résumé Une mutilation et une analgésie acrale sont rapportées chez 13 Epagneul français au Canada. Cette maladie nouvelle présente des caractéristiques similaires frappantes avec les autres syndromes de mutilation acrale ou les neuropathies sensorielles héréditaires rapportées chez les Braque allemand, Pointer et English Springer Spaniels. Les signes cliniques sont initialement notées entre 3.5 et 12 mois d'âge. Les chiens atteints lèchent, mordent et se traumatisent leurs extrémités distales, provoquant des ulcères et une infection bactérienne secondaire. L'auto-amputation des griffes, des doigts et des coussinets est notée dans les cas les plus sévères. Un ou plusieurs pieds peuvent être atteints. Les chiens atteints marchent sur leurs extrémités traumatisées sans montrer de signe de douleur, de boiterie ou d'ataxie. La plupart des chiens ont été euthanasiés quelques jours ou quelques mois après le diagnostic.

Resumen Se describe mutilación acral y analgesia en 13 Spaniels franceses en Canadá. Esta nueva presentación guarda una similitud sorprendente con las características clínicas y las biopsias de los síndromes de mutilación acral o las neuropatías hereditarias sensoriales descritas en el Pointer Alemán de pelo corto, Pointer Inglés, y el Springer Spaniel inglés. Los primeros síntomas clínicos se observan entre los 3,5 y 12 meses de la edad. Los perros afectados se lamen, muerden y se automutilan intensamente las extremidades distales, resultando en úlceras con infección bacteriana secundaria. Se produce auto-amputación de garras, dígitos y almohadillas en casos graves. Pueden afectarse extremidades aisladas o de forma múltiple. Los perros afectados caminaban con patas gravemente mutiladas sin evidencia de dolor, cojera o de ataxia. La mayoría de los perros fueron eutanasiados al cabo de días o meses del diagnóstico.

Zusammenfassung Akrale Mutilation und Analgesie wurde bei 13 Französischen Spaniern in Kanada berichtet. Diese erst kürzlich entdeckte Erkrankung zeigt offenkundige Ähnlichkeit bezüglich klinischer Merkmale und Biopsiefunden mit anderen akralen Mutilationssyndromen oder vererblichen sensorischen Neuropathien wie sie beim Deutsch Kurzhaar, English Pointer und English Springer Spaniel berichtet wurden.