PAPER

Autosomal dominant mutation causing the dorsal ridge predisposes for dermoid sinus in Rhodesian ridgeback dogs

OBJECTIVES: To define the mode of inheritance of the dorsal ridge and investigate if the ridge predisposes to the congenital abnormality dermoid sinus in the Rhodesian ridgeback.

METHODS: Segregation analysis was performed, including 87 litters (n=803) produced in Sweden between 1981 and 2002. Data were corrected to avoid bias in the segregation ratio. Chi-squared analysis was performed including 402 litters (n=3598) for the evaluation of a possible genetic correlation between the ridge and dermoid sinus.

RESULTS: The ridge is inherited in an autosomal dominant mode and predisposes for dermoid sinus. The frequency of ridgeless offspring in the Swedish Rhodesian ridgeback population is estimated to be 5·6 per cent.

CLINICAL SIGNIFICANCE: Rhodesian ridgeback dogs that carry the ridge trait are predisposed to dermoid sinus.

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Journal of Small Animal Practice (2006) **47**, 184–188

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INTRODUCTION

The Rhodesian ridgeback is an African dog breed with a characteristic coat formation denoting the ridge. The origin of the breed is unknown. It has been suggested that several different European dog breeds (Table 1) and the Hottentot hunting dog, an indigenous breed of Africa from which the characteristic ridge originated, have contributed to generate the modern Rhodesian ridgeback breed (Hare 1932, Lutman 1966, Hawley 1984).

The ridge trait is also found in another purebreed, the Thai ridgeback dog. The breed is indigenous to Asia and is also known as the Phu-Quoc dog (Gulf of Siam). To date it is unknown whether the ridge trait originates from the Phu-Quoc dog of Asia (Wegner 1986) or the Hottentot dog of Africa (Hall 2003).

The Rhodesian ridgeback is often associated with a congenital cutaneous defect, dermoid sinus (DS), which occurs with increased frequency in the breed (Salmon Hillbertz 2005). The defect also occurs in the Thai ridgeback dog (N. H. C. Salmon Hillbertz, unpublished data). The genetic relationship between the two ridged breeds remains to be evaluated using the approach described by Parker and others (2004). Helgesen (1991) discussed a historical aspect of the Rhodesian ridgeback, associating ridged dogs with the behavioural hunting traits for which the breed was selected. It has been described that early observers in southern Africa found the ridge to be synonymous with courage, as the ridged dogs had the pre-eminent ability to bay African game, such as lion and treeing leopards (Lutman 1966).

In the original Rhodesian ridgeback standard of the 1920s, the ridge was clearly defined (Hutchinson 1931). The mode of inheritance of the ridge trait has previously been suggested as autosomal recessive (Hawley 1984, Willis 1989, Robinson 1990, Nicholson and Parker 1991). However, these studies were inconclusive since they did not present statistical support for the mode of inheritance.

The aim of the current study was to conclusively define the mode of inheritance of the ridge trait. The analysis was performed using a sufficiently large population material to ensure statistically conclusive results. In Fig 1 ridgeless and ridged siblings are displayed.

Definition of the ridge

To fulfil the modern Rhodesian ridgeback breed standards, the ridge must be distinct, symmetrical and tapering towards the hip bones. The ridge is divided into three main parts (Fig 2): the box, two symmetrical crowns and the tail. The box is also known as the "head" or "swirl" and is the part of the ridge pertaining the crowns. The box may be heart-shaped, square or rounded.

According to Rhodesian ridgeback breed standards, the crowns should be identical

Table 1. Different European dog breeds that have been suggested as contributors to the establishment of the modern Rhodesian ridgeback

Breed	Source
Bloodhound	Hutchinson (1931), Lutman (1966), Murray (1989)
Boar hound	Lutman (1966)
Bulldog	Lutman (1966), Murray (1989), Helgesen (1991)
Deerhound	Lutman (1966)
Foxhound	Murray (1989)
Greyhound	Murray (1989), Helgesen (1991)
Labrador	Hawley (1984)
Mastiff	Lutman (1966), Murray (1989)
Pointer	Hawley (1984), Murray (1989), Helgesen (1991)
Spaniel	Lutman (1966)
Staghound	Murray (1989)
Terrier	Lutman (1966), Murray (1989)

and opposite to each other; thus, the right crown should swirl clockwise and the left, counter-clockwise. Furthermore, the ridge is required to contain only two crowns and the tail should be a minimum of two-thirds of the length of the ridge, even and symmetrical (Lutman 1966, Helgesen 1991). Similar to the hair of the box, the hair of the tail grows in the opposite direction to the hair of the general coat. In the original Rhodesian ridgeback standard, there was no reference to either the crowns or their dorsal position (Hutchinson 1931). The ridge is distinct on a newborn puppy, that is, the anatomical position and morphology do not change from what is displayed at birth (Helgesen 1991).

MATERIALS AND METHODS

The Swedish Rhodesian ridgeback population

Rhodesian ridgeback breeders have been reporting the health status of born litters to the Swedish Rhodesian Ridgeback Club (SRRS) since 1964 (Salmon Hillbertz 2004). According to the SRRS breeding committee, the current population constitutes of approximately 2500 animals (1995 to 2003) (U. Thedin, personal communication).

Data

The litter health status data used for this study were collected by the SRRS from a

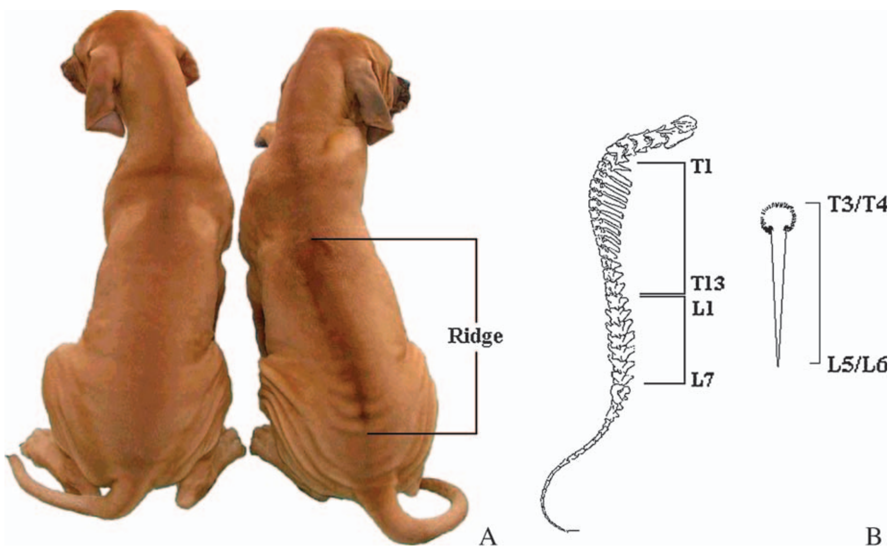

FIG 1. Two Rhodesian ridgeback siblings (A). The left puppy is ridgeless and the right puppy is ridged. The ridge is dorsally located between the thoracic vertebrae (T) T3/T4 and the lumbar vertebrae (L) L5/L6 (B). Photo by R. Hauge. The lateral viewed vertebral column is adapted from Kainer and McCracken (2003)

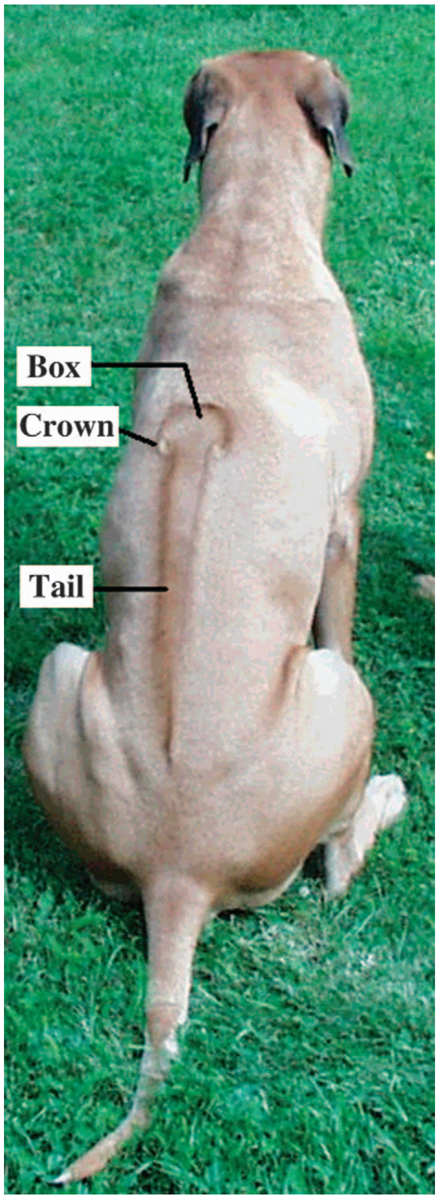

FIG 2. The dorsal ridge in a Rhodesian ridgeback dog. The ridge is divided in three parts, the box, two symmetrical crowns and the tail

total of 402 litters (n=3598) produced between 1981 and 2002. The litters included in the study were exclusively restricted to those where information regarding the number of born offspring in the litters were available and the presence, or absence, of the ridge trait and DS had been recorded. The hypothesis was that the data (Appendix 1) would not deviate from a 3:1 phenotypic ratio (three ridged [*RR* and *Rr*], one ridgeless [*rr*]).

To investigate whether the ridge trait is autosomal dominant and not sex-linked, a four-generation pedigree (U. Thedin, personal communication) was scrutinised. Corrections of expected frequencies were performed as all litters included in the analysis contained one or more ridgeless offspring. The utilised correction formula (Cavalli-Sforza and Bodmer 1971) was $q'=q/(1-p^s)$, where q is the expected frequency of *rr* (0·25), p is the expected frequency of *RR* or *Rr* (1-0·25), q′ is the corrected expected frequency of *rr* and s is the litter size. The segregation analysis was performed to obtain upper and lower estimates of p, by utilising the extended and simplified method of discarding singles (Davie 1979), with the assumption that all families with ridgeless offspring were not included in the data (Nicholas 1987). Further, a chi-squared analysis was performed on all 402 litters (n=3598) to investigate a possible correlation between DS and the ridge trait.

In an effort to investigate whether DS+ ridgeless offspring had been produced in a population other than the Swedish Rhodesian ridgeback population, Joerg Meil, DVM, was consulted. Joerg Meil communicated information from the breeding register of the largest Rhodesian Ridgeback Club in Germany, the Deutsche Züchtergemeinschaft Rhodesian Ridgeback (DZRR), in which two-thirds of German Rhodesian ridgeback litters are registered (approximately 450 litters per year). All Rhodesian ridgeback offspring produced in Germany are examined by trained and qualified personnel.

FIG 3. A four-generation pedigree displaying an autosomal dominant distribution concerning ridged Rhodesian ridgeback offspring. Deceased individuals included in the pedigree did not reach the age of two weeks

RESULTS

No support for a sex-linked distribution of the ridge trait was evident (Fig 3).

Among the records of 402 litters (n=3598), 315 litters (n=2795) showed no evidence of ridgeless offspring. In the remaining 87 litters (n=803), produced by 61 sires and 63 dams, ridgeless offspring were identified. The observed numbers of ridgeless offspring were 202, whereas 601 individuals were defined as phenotypically normal (ridged) according to modern Rhodesian ridgeback breed standards. All 124 parental animals carried a ridge and were thus classified as heterozygotes *Rr*.

Due to the non-randomised selection of litters included in the analysis (n=87), the corrected numbers of ridgeless and ridged offspring show the correct expected frequencies in the selected sample. The results from the segregation analysis (0·77 > P > 0·70; P=0·75) were consistent with an autosomal dominant mode of inheritance (Table 2). Further, a genetic correlation between the ridge and DS was statistically supported (χ^2=12·66 (1 df); P<0·005) (Table 3). No ridgeless DS+ Rhodesian ridgeback offspring had been reported from the German Rhodesian ridgeback population (DZRR) during 2000 to 2003 (201 litters, n=1778) (J. Meil, personal communication).

Based upon reported cases concerning the lack of a dorsal ridge and litter size, the frequency of ridgeless offspring in the Swedish Rhodesian ridgeback population was estimated to be 5·6 per cent (202 ÷ 3598).

DISCUSSION

Availability of the unique Swedish Rhodesian ridgeback register has enabled us to determine that the ridge trait is inherited according to an autosomal dominant mode of inheritance. The autosomal

Table 2. Observed and corrected frequencies for 803 ridged or ridgeless Rhodesian ridgeback dogs

Litter (n)	Sire	Dam	Born	Observed		Expected			
				Ridgeless	Ridged	Uncorrected		Corrected	
						Ridgeless	Ridged	Ridgeless	Ridged
87	61	63	803	202	601	200·75	602·25	217·17	585·83

Data was collected by the Swedish Rhodesian Ridgeback Club during the period 1981 to 2002

Table 3. Observed and expected frequencies for the presence of dermoid sinus (DS) in 3598 ridged or ridgeless Rhodesian ridgeback dogs, produced in Sweden during 1981 to 2002

		DS		
		+	–	Total
Observed	*Rr/RR* (ridged)	201	3195	3396
	rr (ridgeless)	0	202	202
	Total	201	3397	3598
Expected	*Rr/RR* (ridged)	189·72	3206·28	3396
	rr (ridgeless)	11·28	190·72	202
	Total	201	3397	3598

dominant inheritance also corroborates with the distribution over generations of produced ridgeless offspring, shown in Fig 3. The provided association between the congenital skin abnormality DS and the ridge is, to the author's knowledge, the first study to show a statistically supported genetic correlation between these traits, as no ridgeless individuals affected by DS were produced between 1981 and 2002 in Sweden. These results corroborate with the information received from the DZRR. The data concerning *rr* and DS appearances were reported by breeders to the SRRS and therefore the results entirely rely upon the breeders information. Further, it is undetermined whether Swedish breeders from 1981 to 2002 examined all stillborn or euthanased offspring for DS. Therefore, an uncertainty in the absolute numbers of DS+ offspring exists.

The causative mutation (*R*) leading to the existence of the dorsal ridge in this breed is currently unidentified, and there is a lack of knowledge concerning whether the trait originated from the ridged Rhodesian or Thai ridgeback dogs. However, a recent study of the genetic diversity between a large number of dog breeds (Parker and others 2004) may supply the necessary tools regarding evaluating the genetic relationship between dog breeds carrying the *R* mutation. The present study provides knowledge that could aid in the identification of such mutation. Further analysis will allow us to elucidate the genetics underlying the two traits.

Acknowledgements

A special gratitude to the Swedish Rhodesian ridgeback breeders and the SRRS, who have made this study possible. Many thanks to Jörg Meil (Germany), Janet Murray (Australia) and Ulla Thedin (Sweden) for personal communications, Ronny Hauge (Norway) for photographs, and to Dr Carl-Gustaf Thulin, Professor Leif Andersson and Professor Per-Erik Sundgren for helpful discussions and comments on the manuscript. Funding was provided by the Swedish Kennel Club research foundation and the SRRS.

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Appendix 1. Data derived from 87 litters, where ridgeless offspring were identified from 1981 to 2002

						Expected			
				Observed		Uncorrected		Corrected	
Litter No.	Sire	Dam	Born	Ridgeless	Ridged	Ridgeless	Ridged	Ridgeless	Ridged
1	Y1	X1	11	3	8	2·75	8·25	2·87	8·13
2	Y2	X2	4	3	1	1·00	3·00	1·46	2.54
3	Y3	X3	5	2	3	1·25	3·75	1·64	3·36
4	Y4	X4	9	2	7	2·25	6·75	2·43	6·57
5	Y5	X5	11	3	8	2·75	8·25	2·87	8·13
6	Y6	X6	17	3	14	4·25	12·75	4·28	12·72
7	Y7	X4	7	2	5	1·75	5·25	2·02	4·98
8	Y8	X7	12	1	11	3·00	9·00	3·10	8·90
9	Y9	X8	9	3	6	2·25	6·75	2·43	6·57
10	Y9	X9	7	1	6	1·75	5·25	2·02	4·98
11	Y10	X10	8	3	5	2·00	6·00	2·22	5·78

(*continued*)

Appendix 1. (continued)

						Expected			
				Observed		Uncorrected		Corrected	
Litter No.	Sire	Dam	Born	Ridgeless	Ridged	Ridgeless	Ridged	Ridgeless	Ridged
12	Y11	X11	10	2	8	2·50	7·50	2·65	7·35
13	Y11	X12	6	2	4	1·50	4·50	1·82	4·18
14	Y12	X13	9	1	8	2·25	6·75	2·43	6·57
15	Y13	X14	11	1	10	2·75	8·25	2·87	8·13
16	Y13	X15	9	3	6	2·25	6·75	2·43	6·57
17	Y14	X16	6	1	5	1·50	4·50	1·82	4·18
18	Y15	X17	8	1	7	2·00	6·00	2·22	5·78
19	Y16	X18	8	1	7	2·00	6·00	2·22	5·78
20	Y17	X19	10	5	5	2·50	7·50	2·65	7·35
21	Y18	X6	11	4	7	2·75	8·25	2·87	8·13
22	Y19	X20	10	3	7	2·50	7·50	2·65	7·35
23	Y20	X21	8	3	5	2·00	6·00	2·22	5·78
24	Y21	X22	10	4	6	2·50	7·50	2·65	7·35
25	Y21	X23	12	4	8	3·00	9·00	3·10	8·90
26	Y22	X24	9	2	7	2·25	6·75	2·43	6·57
27	Y23	X25	9	3	6	2·25	6·75	2·43	6·57
28	Y23	X26	8	3	5	2·00	6·00	2·22	5·78
29	Y23	X27	9	2	7	2·25	6·75	2·43	6·57
30	Y23	X28	8	4	4	2·00	6·00	2·22	5·78
31	Y24	X29	3	1	2	0·75	2·25	1·30	1·70
32	Y24	X30	11	5	6	2·75	8·25	2·87	8·13
33	Y25	X31	9	2	7	2·25	6·75	2·43	6·57
34	Y25	X32	9	3	6	2·25	6·75	2·43	6·57
35	Y26	X26	6	1	5	1·50	4·50	1·82	4·18
36	Y26	X33	12	6	6	3·00	9·00	3·10	8·90
37	Y27	X34	11	5	6	2·75	8·25	2·87	8·13
38	Y28	X19	11	1	10	2·75	8·25	2·87	8·13
39	Y29	X35	9	5	4	2·25	6·75	2·43	6·57
40	Y29	X36	8	1	7	2·00	6·00	2·22	5·78
41	Y30	X37	10	2	8	2·50	7·50	2·65	7·35
42	Y30	X38	7	1	6	1·75	5·25	2·02	4·98
43	Y31	X37	11	6	5	2·75	8·25	2·87	8·13
44	Y32	X39	9	1	8	2·25	6·75	2·43	6·57
45	Y32	X38	10	3	7	2·50	7·50	2·65	7·35
46	Y32	X40	10	3	7	2·50	7·50	2·65	7·35
47	Y32	X41	3	2	1	0·75	2·25	1·30	1·70
48	Y33	X42	9	1	8	2·25	6·75	2·43	6·57
49	Y34	X19	8	1	7	2·00	6·00	2·22	5·78
50	Y35	X43	7	2	5	1·75	5·25	2·02	4·98
51	Y35	X44	10	2	8	2·50	7·50	2·65	7·35
52	Y36	X45	9	3	6	2·25	6·75	2·43	6·57
53	Y37	X16	10	1	9	2·50	7·50	2·65	7·35
54	Y38	X46	10	1	9	2·50	7·50	2·65	7·35
55	Y39	X47	9	2	7	2·25	6·75	2·43	6·57
56	Y40	X2	9	1	8	2·25	6·75	2·43	6·57
57	Y40	X48	9	1	8	2·25	6·75	2·43	6·57
58	Y40	X49	8	2	6	2·00	6·00	2·22	5·78
59	Y40	X48	10	1	9	2·50	7·50	2·65	7·35
60	Y40	X50	9	2	7	2·25	6·75	2·43	6·57
61	Y41	X51	14	1	13	3·50	10·50	3·56	10·44
62	Y42	X52	10	1	9	2·50	7·50	2·65	7·35
63	Y43	X53	9	1	8	2·25	6·75	2·43	6·57
64	Y44	X54	8	2	6	2·00	6·00	2·22	5·78
65	Y45	X55	9	4	5	2·25	6·75	2·43	6·57
66	Y45	X56	10	3	7	2·50	7·50	2·65	7·35
67	Y46	X57	11	1	10	2·75	8·25	2·87	8·13
68	Y47	X58	9	4	5	2·25	6·75	2·43	6·57
69	Y48	X59	11	3	8	2·75	8·25	2·87	8·13
70	Y49	X60	11	5	6	2·75	8·25	2·87	8·13
71	Y50	X37	9	2	7	2·25	6·75	2·43	6·57
72	Y51	X6	12	1	11	3·00	9·00	3·10	8·90
73	Y52	X13	11	1	10	2·75	8·25	2·87	8·13
74	Y52	X29	11	4	7	2·75	8·25	2·87	8·13
75	Y52	X61	9	1	8	2·25	6·75	2·43	6·57
76	Y53	X21	6	1	5	1·50	4·50	1·82	4·18
77	Y54	X62	10	1	9	2·50	7·50	2·65	7·35
78	Y55	X29	10	1	9	2·50	7·50	2·65	7·35
79	Y55	X14	10	3	7	2·50	7·50	2·65	7·35
80	Y55	X52	11	3	8	2·75	8·25	2·87	8·13
81	Y56	X21	8	2	6	2·00	6·00	2·22	5·78
82	Y57	X1	12	3	9	3·00	9·00	3·10	8·90
83	Y58	X63	10	1	9	2·50	7·50	2·65	7·35
84	Y58	X16	10	4	6	2·50	7·50	2·65	7·35
85	Y59	X29	7	3	4	1·75	5·25	2·02	4·98
86	Y60	X44	11	2	9	2·75	8·25	2·87	8·13
87	Y61	X38	7	1	6	1·75	5·25	2·02	4·98
87	**61**	**63**	**803**	**202**	**601**	**200·75**	**602·25**	**217·17**	**585·83**