HEREDITARY CONGENITAL ATAXIA IN HOLSTEIN-FRIESIAN CALVES

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SUMMARY

A total of eight calves showing spastic symptoms was found in two related herds of Holstein-Friesian cattle over a 2-yr. period. The symptoms were those of incoordination or ataxia, a failure of synergic muscle groups to act harmoniously. The symptoms were first apparent at about 6 wk. of age and were similar in all cases. The disease is not sex-linked, since calves of both sexes were affected. An analysis of the pedigrees of the calves revealed one common male ancestor. This bull appeared in the pedigrees of both the sire and dam of all the affected calves. The ataxia is postulated to be inherited as an autosomal recessive. The occurrence of ataxic calves ceased when bulls known to be carriers were removed from the herd. The anatomical basis for the condition was neuronal degeneration and cerebellar hypoplasia.

The economic importance of lethals in cattle can be great. The average cow produces only a few offspring in a lifetime. Lethals preventing the perpetuation of a particular line of breeding containing genes of desired characters could result in great economic loss. The total loss due to inherited lethals is not known. However, they would constitute a portion of the calves born dead. This number has been tabulated by Linn (7) at between 5 and 6% in approximately 25,000 calves born to Kansas D.H.I.A. cows.

A realization of the importance of the problem of inherited lethals in dairy cattle has been accentuated by the rapid growth of artificial breeding. Gilmore (3) lists 28 lethal characters in cattle, two of which are congenital spasms and cerebellar hypoplasia.

Saunders et al. (8) reported nervous symptoms in 23 calves born in a purebred Jersey herd over a 15-yr. period. The clinical symptoms were those of incoordination. The symptoms were present at birth in some of the calves and appeared during the first week or two of life in the others. Although such calves could nurse and had good appetites, all died within a few weeks. The anatomical basis for the symptoms was an anomaly of the cerebellum and midbrain.

Congenital spasms were reported by Gregory et al. (4) to occur as a recessive lethal condition in a Jersey herd. Affected calves exhibited intermittent spasmodic movements of the head and neck, usually in a vertical plane. When the animals were forced to stand, spasms in both front and hind legs hampered standing or walking. Aside from the spasms, such calves, up to a few days of age, appeared

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to be normal and vigorous and to have good appetites. They died, however, within a few weeks after birth.

A familial cerebellar ataxia in purebred Hereford calves in England was reported by Innes et al. (6). The syndrome was apparent at birth and was clinically similar to that reported in Jersey calves by Saunders et al. (8). The cerebellum of affected calves was abnormally small, and was tough in consistency. The lesion was interpreted histologically as a cortical hypoplasia. In two of these herds, replacement of the bulls resulted in a cessation of the disease. Hereford bulls that had sired ataxic calves from Hereford cows sired only normal offspring from Shorthorn cows. The disease was thought to be due to a lethal recessive gene that was restricted to the Hereford breed.

Congenital spasms were reported by Cole (2) as a recessive lethal condition in Holstein-Friesian calves. A 3-mo-old Holstein-Friesian bull calf with congenital cerebellar hypoplasia was found in the Denver, Colorado, Stockyards by Anderson and Davis (1). Congenital hereditary cerebellar ataxia has been observed in various breeds of sheep in Great Britain and Canada by Innes and MacNaughton (5).

Some diseases with comparable clinical symptoms have been presented in this review of literature for purpose of comparison. However, none of the calves discussed in the reviews were so long in developing the disease or survived so long afterwards as the calves in this study.

MATERIALS AND METHODS

Eight calves with extreme spasmodic movements were observed (over a 2-yr. period) in two related herds of Holstein-Friesian cattle. The eight calves, six females and two males, of which one male and one female were full brother and sister, all exhibited the same clinical symptoms. The age at which the symptoms were first observed varied from 6 wk. to 5 mo. The ataxia was barely noticeable when first observed and grew progressively worse until the calves were destroyed. Since almost all of the bull calves from the two herds were sold at three days of age, it is not known how many of these calves would have developed the disease.

The clinical symptoms for all the affected calves were those of incoordination or ataxia, a failure of synergic muscle groups to act harmoniously. The calves exhibited intermittent spasmodic movements of the head and neck, usually in a vertical plane. When the animals were forced to stand, spasms in both front and hind legs hampered standing or walking. Animals stood with all four legs braced in a straddled position and, when attempting to move, showed marked incoordination (Figure 1). An attempt to walk resulted in a fall, usually backward (Figure 2). Noise and shock neither initiated nor intensified the spasms. Aside from the spasms, such calves appeared to be normal and vigorous and to have good appetites.

RESULTS

Five calves, four female and one male, that had become spastic and developed ataxia were autopsied at 6–13 mo. of age by the Department of Veterinary
Pathology, State College of Washington. The case history and autopsy results of the five calves were so similar that there is little doubt that the calves had the same disease. Since the cases were so similar, an autopsy report is given for the oldest calf observed.

This calf was born in February and the spastic condition first noted in July. The calf, until that time, had appeared to be normal, with a good appetite, and had a normal rate of gain. The spastic condition developed into ataxia in about 6 wk. The ataxia progressed until the calf could no longer stand after being helped to its feet. Autopsy was performed in March at 13 mo. of age. The results of the autopsy were: main thoracic and abdominal organs normal with the exception of the kidney, where small, pale, wedge-shaped areas were
observed in the cortex. Microscopically, these were found to be focal areas of interstitial nephritis. The brain appeared small and was found to weigh 301.3 g. The brain of a normal calf of nearly the same size examined on the same day weighed 450 g. The convolutions of the cerebellum appeared small and very closely packed together and, in at least two instances, they projected upward into the dura in a papillary fashion. The dura appeared to be thickened over the area of the cerebellum and was found to have a slight degree of fibrosis.

The microscopic examination of the brain revealed a mild neuronal degeneration in the thalamus, the cerebellum, and the cerebrum. There was moderate satellitosis and gliosis in all areas examined, and the cerebellum showed a conspicuous degeneration of the Purkinje cells, with a reduction in the number of cells in the granular layer. There was a moderate diffused gliosis and a slight neuronal degeneration in the medulla. Sections of the spinal cord appeared normal.

There appeared to be some distortion of the cranial cavity, with a reduction in the space available for the cerebellum and the medulla. The tentorium, which tends to separate the medulla and cerebellum from the cerebrum, was very small and poorly developed. There is some possibility that the failure of the development of the bones of the cranial cavity, or some distortion in the direction of development of the bones, may have caused increased pressure on the brain and interfered with its proper development.

The pedigrees of the affected calves were studied, to determine whether or not the condition could have been inherited. One male was found to appear in the pedigree of both the sire and dam of all the affected calves (Figure 3). Calves 1 and 2 were full brother and sister. They trace back to this common male ancestor five generations through their sire's pedigree and four generations through their dam's pedigree. Calf 3 is four generations from this ancestor through both the sire's and the dam's pedigree. Calves 4 and 8 are four generations away through the sire's pedigree and five generations away through the

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**Fig. 3. Pedigree showing inheritance of congenital ataxia in Holstein-Friesian calves.**
dam’s pedigree. Calves 5, 6, and 7 are five generations away through both their sires’ and dams’ pedigrees. Although all the animals listed as known carriers in Figure 3 did not produce ataxic calves, they are in the direct line of descent from the one common ancestor. Therefore, it was assumed that they were carrier animals.

None of the parents of the ataxic calves showed any of the nervous symptoms. Since the affected offspring were born from phenotypically normal parents, the disease, if inherited, would have to be due to a recessive gene. As both sexes were included among the affected calves, the condition could not be sex-linked. No graduation in the symptoms was observed. The calves either had all the symptoms or none, thus eliminating multiple factors, unless the disease was due to threshold-type inheritance. It was postulated that the ataxia was conditioned by an autosomal recessive gene. The breeding of heterozygous carriers would then result in affected calves whenever a calf was homozygous for this disease.

A planned inbreeding program was carried out in which a known carrier bull was bred to his daughters and daughters of his half-brother, including three known carrier females. One of three resulting calves was affected.

The occurrence of ataxic calves has ceased since males and females known to be carriers of the disease have been removed from the herds.

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REFERENCES