Short Communications

Congenital reflex myoclonus in two Merino cross lambs in South Africa

D. J. C. Blignaut, D. E. Holm, R. Leask, N. Stander, J. C. A. Steyl

REFLEX myoclonic conditions have been reported in mice, labrador retriever dogs (March and others 1993), Peruvian Paso horses (Gundlach and others 1993) and Hereford calves (Harper and others 1986). The primary cause of myoclonus in these reports was consistent with a deficit in glycine receptors in the CNS (Pierce and others 2001). Similar conditions based on clinical findings and the lack of pathological lesions have not been reported in sheep.

Two three-day-old, female Dohne Merino × Merino lambs from different ewes (assigned lamb 1 and lamb 2 for identification purposes) were presented with a complaint of being unable to stand, having tremors and showing signs of stiffness. The flock was managed as a multisire breeding system. Annual vaccinations were done against conditions such as pulpy kidney, bluetongue, blue udder and Chlamydophila. Only these lambs were affected in the flock. Both lambs had received sufficient colostrum from their respective dams shortly after they were born.

A complete clinical examination, neurological examination, blood smear for light microscopy evaluation, urinalysis and faecal analysis were performed. Both lambs appeared to be alert, could lift their heads and had no visual or auditory abnormalities. The lambs were able to suckle normally when assisted, but only when left in lateral recumbency. The lambs were recumbent and unable to rise or stand on their own and showed limited voluntary movement of the fore and hindlimbs. Attempts to make the lambs stand only induced the typical whole body rigidity with the head and neck extended and characteristic hindlimb adduction to crossing over in a rigid extension manner (as seen by Blood and Gay 1971, Harper and others 1986, Windsor, personal communication). These myoclonic episodes were consistently characterised by rigidity of the body, extensor spasms, clearly palpable tachycardia and temporary apnoea. On return to lateral recumbency, both lambs resumed the initial posture, spasms ceased and normal respiration resumed after a short period of open mouth breathing. Sound stimuli in the form of sudden loud noises and sensory stimuli in the form of touch produced a milder response. Repetition of a sound stimulus (such as clapping the hands) lessened the intensity of the response until there was no response. Withdrawal reflexes in the fore and hindlimbs were present, indicating entire spinal cord segments from C6 to T2 in the forelimbs and the radial nerve and spinal cord segments L5 to S1 in the hindlimbs and sciatic nerve (as observed by Constable 2004). Correction of the abnormal position during knuckling of the distal limbs seemed to be delayed, but it is believed that the whole body rigidity during manipulation obscured this procedure. Lamb 2 could stand for a short period with assistance after repeated attempts and was able to support its own weight while the whole body rigidity subsided momentarily, until it was stimulated again.

FIG 1: Lamb showing myoclonic whole body rigidity with cross-over extension of the hindlimbs

Whole blood in EDTA and serum were collected for haematology and serum concentrations of sodium, potassium, urea, creatinine, total calcium, magnesium, inorganic phosphorous, aspartate transferase, creatine kinase and bile acids were measured. Abnormalities detected are summarised in Table 1. All other clinical chemistry results for both lambs were within the normal reference ranges. The mildly elevated serum creatine kinase, serum urea and blood platelet count were considered to be of less significance to the diagnosis of this particular case.

Cisternal cerebrospinal fluid samples were collected and analysed. The lambs were sedated with 0.5 mg midazolam (Dormicum; Roche Products) and 20 mg ketamine (Ketamine-Fresenius; Bodene). Gross appearance of both samples was clear and colourless. Specific gravity, red cell count, white cell count and protein concentration were within normal reference ranges (Scott 2010). Extended ventrodorsal pelvic radiographs of each lamb were taken. Coxofemoral abnormalities were ruled out. The consistent hip joint lesions that were seen in polled Hereford calves with inherited congenital myoclonus (ICM) were not observed radiographically. It was suggested that the hip joint lesions seen in the calves with ICM were secondary as a result of severe myoclonic contractions of the pelvic adductor muscles seen by extensor crossing of the hindlimbs (Harper and others 1986). It is possible that these sheep are traditionally selected for wool and not muscle mass, therefore, the possibility of having a relatively smaller adductor muscle mass might influence the frequency at which these distinctive hip joint lesions were seen.

CT images of the neurocranium of lamb 1 were taken to investigate any gross structural abnormalities and cerebellar hypoplasia antemortem. There were no visible gross structural abnormalities and the cerebellum was of normal size.

Lamb 1 was treated with 10.2 mg vitamin E acetate and 0.4 mg selenium (VitESe Injection; Kyron Laboratories). The neurological signs present and lack of improvement in clinical signs after treatment excluded any deficiencies in these elements and were not investigated further.

Veterinary Record (2011) 169, 684b doi: 10.1136/vr.d5812

D. J. C. Blignaut, BVSc,
D. E. Holm, BVSc, MSc (Vet), MRCVS,
R. Leask, BSc (Agric), BVSc,
MMedVet (CaprOv),
Department of Production Animal Studies,
N. Stander, BVSc, MMedVet (DiagIm),
Department of Companion Animal Clinical Studies,
J. C. A. Steyl, BVSc,
Section of Pathology, University of Pretoria, Private Bag X04, Onderstepoort, 0110, South Africa

E-mail for correspondence: dawie.blignaut@up.ac.za

Provenance: not commissioned; externally peer reviewed

Published Online First October 6, 2011

TABLE 1: Comparative abnormalities observed between lamb 1 and lamb 2

	Lamb 1	Lamb 2	Reference range
Identification number	05/218	PU03204	
Severity of myoclonic spasms	More severe	Less severe	
CT findings	Normal	Not done	
Response to treatment with vitamin E/selenium	No response	Not treated	
Serum creatine kinase (U/l)	110	61	12-51
Serum urea (mmol/l)	7.50	6.20	2.65-2.64
Serum creatinine (µmol/l)	73	40	44-150
Blood neutrophil count (× 10^9/l)	4.35	5.71	0.40-5.00
Blood platelet count (× 10^9/l)	1505	2317	250-750

After a period of two weeks, the lambs were humanely euthanased with intravenous overdose of pentobarbitone and submitted for postmortem examination. No significant pathological lesions could be demonstrated in any tissues on routine macroscopic or histological examination. There were no abnormalities in the hip joints of both lambs. Long-term nursing of the recumbent lambs was complicated by dermal pressure injuries. The lack of pathological lesions at macroscopic and histological level ruled out a number of differential diagnoses affecting the neuromuscular and skeletal systems in lambs.

A specific aetiology for the condition could not be demonstrated with routine diagnostic procedures. A presumptive diagnosis of congenital reflex myoclonus was made. The diagnosis is based on clinical similarities between other animal models previously studied that showed reflex myoclonus upon stimulation. To investigate the possibility of a molecular neuromuscular cause, further specific genetic and immunohistochemistry analysis would be required to study the condition seen in these Merino cross lambs.

Acknowledgements

The authors acknowledge the Onderstepoort Veterinary Academic Hospital, the Department of Production Animal Studies and the Veterinary Genetics Laboratory, University of Pretoria, for financial support as well as Kate Moseley and other final-year veterinary students who cared for the lambs during hospitalisation. They also thank Dr C.K. Harper (Veterinary Genetics Laboratory) for her efforts with genetic analysis.

References

BLOOD, C. D. & GAY, C. C. (1971) Hereditary neuraxial oedema of calves. *Australian Veterinary Journal* **47,** 520

CONSTABLE, P. D. (2004) Clinical examination of the ruminant nervous system. *Veterinary Clinics of North America: Food Animal Practice* **20,** 185-214

GUNDLACH, A. L., KORTZ, G., BURAZIN, T. C., MADIGAN, J. & HIGGINS, R. J. (1993) Deficit of inhibitory glycine receptors in spinal cord from Peruvian Pasos: evidence for an equine form of inherited myoclonus. *Brain Research* **628,** 263-270

HARPER, P. A., HEALY, P. J. & DENNIS, J. A. (1986) Inherited congenital myoclonus of polled Hereford calves (so-called neuraxial oedema): a clinical, pathological and biochemical study. *Veterinary Record* **119,** 59-62

MARCH, P. A., KNOWLES, K. & THALHAMMER, J. G. (1993) Reflex myoclonus in two Labrador Retriever littermates: a clinical, electrophysiological and pathological study. *Progress in Veterinary Neurology* **4,** 19-24

PIERCE, K. D., HANDFORD, C. A., MORRIS, R., VAFA, B., DENNIS, J., HEALY, P. J. & SCHOFIELD, P. R. (2001) A nonsense mutation in the α1 subunit of the inhibitory glycine receptor associated with bovine myoclonus. *Molecular and Cellular Neuroscience* **17,** 354-363

SCOTT, P. R. (2010) Cerebrospinal fluid collection and analysis in suspected sheep neurological disease. *Small Ruminant Research* doi: 10.1016/j.smallrumres.2010.04.009