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Congenital contractural arachnodactyly ('fawn calf syndrome') in Angus cattle

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CA or 'fawn calf syndrome' – what is it?

Congenital contractural arachnodactyly (CA), also known as "fawn calf" syndrome (FCS), is a heritable disease of newborn Angus and Angus-derived cattle caused by an error in the DNA genetic code transmitted from parents to their progeny. Calves with CA are visibly abnormal at birth (Fig. 1) but are usually born alive. Most survive to adulthood. CA is a developmental defect involving reduced elasticity of the connective tissue of muscles, first identified in Victoria in 1998 but now reported in many countries. The Angus Society of Australia lists FCS as a "Category 1 Genetic Condition" (Regulation. 6.24) (www.angusaustralia.com.au/Regos_Regulations.htm#6). Animals known to express any Category 1 Genetic Condition are ineligible for registration with the Society. Known carriers of Category 1 defects are identified as such in the Australian Angus Database (www.angusaustralia.com.au/DBS.htm).

Diagnosis of CA in the newborn calf

Diagnosis of CA is made by physical examination of the live newborn calf, preferably within 24 hours of birth and preferably by a veterinarian or breeder familiar with CA. Although there is some variation in severity between cases, CA is easily diagnosed in the live newborn calf. Both sexes can be affected. To make the diagnosis in a newborn calf, it is necessary that *all* of the following are observed:

- Congenital proximal limb contracture;
- Congenital distal limb hyperextension;
- Congenital kyphosis; *and*
- Significant post-natal improvement in these clinical signs as the calf grows and matures.

(These clinical signs are explained in detail below.)

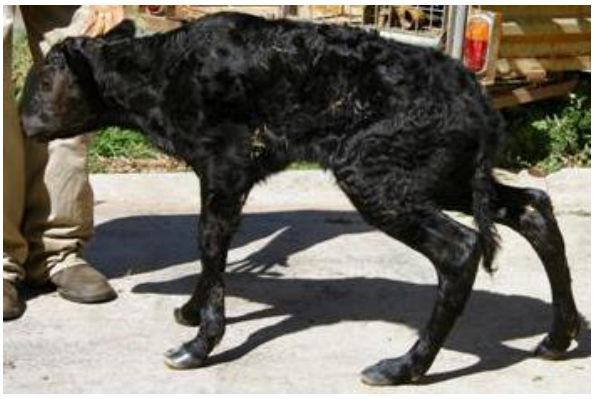

Figure 1. Congenital contractural arachnodactyly (CA) in a 1 day old Angus calf. Note the angulation of stifle and hock joints due to muscle contracture and hyperextension of loose-jointed fetlocks (calf is 'down on its pasterns'). Typical "fawn calf".

In less severe cases, kyphosis (upwards arching of the spine) and fetlock hyperextension (see Fig. 1) at birth often resolve almost completely within a week or so after birth (see Fig. 3). Hyperextension of the fetlocks results from 'loose jointedness', present throughout the skeleton but difficult to detect in the proximal joints affected by contracture of muscles.

Muscle contracture (loss of extensibility) and the resulting inability to straighten the proximal (upper limb) joints takes longer to resolve, but can be difficult to detect by four weeks of age in mild cases. In severe cases, kyphosis and limb contracture remain to adulthood, despite improvement.

CA has not been reported in any breed of cattle other than Angus to date. No other syndrome with this combination of clinical signs at birth has been reported in any breed, although a related disorder (Marfan's syndrome) has been reported in newborn Limousins. Importantly, the post-natal improvement seen in CA has not been reported in any other congenital syndrome of cattle with proximal contractures and distal hyperextension.

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Diagnosis of CA is not difficult, but requires careful physical examination of the newborn calf and then subsequent observation to detect the improvement.

Additional clinical signs are usually present in newborn CA cases but can be more difficult to appreciate, especially in mild cases. These include:

- Unusually long leg bones resulting in abnormally tall stature and higher frame scores;
- Loose-jointedness in other joints (i.e. in addition to the distal limb joints)
- Poor muscling; and
- 'Knock-knees' (valgus deformity).

Proximal contracture (i.e. upper limb contracture) is detected by placing the live calf on its side and pulling down and backwards on a hindfoot while holding the pelvis in place with the other hand if necessary, thereby revealing any reduction in the normal range of angular movement of the proximal joints. In CA, the upper limb joints cannot be straightened out (extended) to the normal extent. This is most obvious in the stifle and hock joints, but also affects the hips. This diagnostic procedure for proximal contracture is important, to demonstrate that the increased angulation of the proximal joints seen when the suspect CA calf is standing (Fig. 1) is really associated with *reduction* of normal angular joint movement due to the muscle contracture and is not just increased angular joint movement as seen in calves with other congenital syndromes involving generalised loose-jointedness. Hindlimb proximal contracture causes some severe CA cases to adopt a "bunny-hopping" gait when trying to run.

Figure 2. Diagnosis of CA. Proximal contracture of hindlimb is detected by pulling downwards and backwards on the hindfoot to demonstrate that the stifle, hock and hip joints cannot be straightened out (extended) to the normal extent.

Distal hyperextension can be recognised by observing that the CA calf is "down on its pasterns" when standing (ie hyperextended fetlocks) (Fig. 1). The underlying distal joint laxity is demonstrated with the calf on its side by an increased range of angular passive movement in the fetlock joints.

Generalised hyperlaxity or 'loose-jointedness' in CA is demonstrated by an increased range of passive movement in joints throughout the skeleton, but this can be difficult to appreciate for those who do not regularly undertake physical examinations of calves. **Scoliosis** is present in severe CA cases, seen as twisting of the spine (deviation from the normal straight line of the vertebrae when looking from above). **Muscle hypoplasia** (due to poor muscle development) is most evident in the hindlimbs, particularly in the major muscles behind the stifle. Intermittent dislocation of the patella in one or both stifles while walking is also common in severe CA cases.

Post-mortem examination is not an alternative to a proper examination of the live calf for diagnosis of CA and usually adds little if anything to a diagnosis made by examination of the live calf. A DNA test for CA is now under development in the USA. but at this time no laboratory test is available to assist or confirm the diagnosis of CA.

Diagnosis of CA in older calves

Diagnosis of CA is more difficult in older calves, particularly in cases that were only mildly affected at birth. However, if there is demonstrable excess elongation of the leg bones in an older calf with residual proximal hindlimb contracture, a diagnosis of CA is possible despite resolution of the fetlock hyperextension and kyphosis present at birth. To make a reliable diagnosis of CA in an older calf, age-matched normal siblings must be available for comparison of frame scores (body height) in order to be confident that excess elongation of the long bones is really present. A diagnosis of CA should not be made unless proximal contracture *and* excess long bone growth are both present. The latter can be difficult to demonstrate in field cases.

Figure 3. CA affected calf at 6 days of age (same calf as Fig 1.) Note the significant improvement in kyphosis and distal hyperlaxity. Longer than normal leg bones and poor muscling are also evident.

Why is CA important?

Without human intervention up to 20% of CA calves will die soon after birth, simply because they are unable to stand up to suckle. Those CA calves that survive are prone to joint dislocations and the early onset of degenerative arthritis as a result of their persistent joint laxity. Although CA is a less severe disease than lethal genetic defects of Angus calves such as α-mannosidosis (MA), arthrogryposis multiplex (AM) and neuropathic hydrocephalus (NH), CA is nonetheless a significant cause of calf wastage in herds where prevalence of the CA mutation in the cow herd is high and carrier bulls are being used.

Figure 4. CA calf, 20 months (left side) with age matched sibling that was normal at birth. The affected calf is taller (higher frame score) with poor muscle development. Same case as Figs.1 and 3.

How does CA spread between herds?

CA is a recessive genetic defect. This means the disease is only seen in a calf that has inherited two copies of the CA mutation, one from each parent. Both parents of any CA affected calf must therefore necessarily be 'carriers' of the mutation. Calves that inherit only one copy of the mutation appear normal but are nonetheless genetic 'carriers' of the mutation and transmit the mutation to 50% of their own progeny.

When CA carriers are mated to other CA carriers, only 25% of the resulting progeny will be affected by the CA disease. Half of the resulting progeny will be apparently normal but actually carriers of the mutation (i.e. the same status as their parents) with the remaining 25% appearing normal and being non-carriers.

CA is usually introduced into a herd by introduction of a CA carrier bull, but can be introduced by CA carrier cows and even by the introduction of an animal that was itself a mildly affected "fawn calf" (ie. a CA affected calf) at birth but not recognised as such in adulthood.

Figure 5. CA, 6 year old cow. Same case as Figs. 1, 3 and 4, shown here with her 9 month old CA affected calf sired by the affected bull in Fig. 8.

What are the risks from FCS for producers?

In Australia, CA has only been detected in the descendants of *Freestate Barbara 871 of Kaf*, a US Angus cow born in Indiana in 1978. Pedigree analysis is therefore a useful adjunct in diagnosis of CA. However, almost half the Angus cattle in Australia are descendents of this US Angus cow and hence are potential CA carriers, even though in reality most of these descendants will not actually be carriers. It is now estimated that less than 5% of Angus cattle in Australia will be CA carriers.

At the moment it is not possible to distinguish those animals that are genetic carriers without showing any signs of the disease from those animals that are truly non-carriers. A DNA test is needed for this.

Many Angus bulls descended from *Freestate Barbara* were used in Australia in the last 20 years and identified as CA carriers when they sired CA affected calves. The risk of any descendent of *Freestate Barbara* being a carrier itself is higher where that descendent has a known carrier close up in its pedigree, for example as a parent (50% risk) or grandparent (25% risk). Descendents with pedigrees in which the closest identified CA carrier is a more distant ancestor are a lower risk.

Angus sires known to be CA carriers

- Rambo 465T of JRS
- Te Mania Kelp K207
- Bon View Bando 598
- SAF 598 Bando 5175
- Boyd On Target 1083

Te Mania Kelp is an Australian bred sire. The other bulls are US bred sires with imported semen. By necessary implication from the lines of descent in pedigrees of CA cases, *Premier Independence KN*, *Shadymere Freedom* and *O'Neill's Renovator* are 'suspect' carriers of CA that may have transmitted the CA mutation to 50% of their progeny.

Figure 6 'Knock-knees' (valgus deformity) in a newborn CA affected calf. Typical 'fawn calf'.

When should you suspect that you have a CA calf ('fawn calf') in your herd?

If you have a newborn Angus or Angus-cross calf in your herd that meets the diagnostic criteria set out on the first page of this document, you should suspect CA. If you are not confident in making the diagnosis, seek assistance from a veterinarian or experienced Angus breeder. Remember that a physical examination as soon as possible after birth is often critical for a reliable diagnosis. ***Keep this document to show to your veterinarian.***

What should you do if you suspect that you have a CA calf ('fawn calf') in your herd?

Further cases can be avoided by ensuring that carrier cows are not joined to carrier bulls. Pedigree analysis may be necessary. If the parents of an affected calf are rejoined, there is a 25% recurrence risk that future progeny will be born with CA.

If you have a number of genetically related cows in your herd that may be CA carriers based on a pedigree analysis, you may decide to use bulls from a different bloodline to reduce the risk of CA calves.

If you are a registered Angus breeder you should notify the Angus Society of Australia of any CA case in your herd. If you have a CA affected calf that is not a descendent of any of the bulls named above, you should certainly always report such a case.

If you are a commercial breeder with fawn calves in your herd, further cases can be avoided by ensuring the bulls you use are a low risk for CA (i.e. do not have a known carrier close up in their pedigree). Registered Angus bulls with no known carrier in their pedigree are a low risk. Advice can be obtained from Industry & Investment, NSW.

Are there any other syndromes that can easily be confused with CA?

If careful examination of the live calf is undertaken as described in this document, there should be no confusion between CA and other syndromes.

Why is 'fawn calf syndrome' now called congenital contractural arachnodactyly?

The term 'fawn calf' has caused confusion because people expect these calves to be fawn in colour but they were actually called 'fawn calves' due to their appearance being similar to that of a newborn deer. The clinical signs in 'fawn calves' are, however, similar to a well-defined syndrome in other species including humans that has long been known as *congenital contractural arachnodactyly* (CCA or CA). ('Arachnodactyly' just means 'spider fingers' and refers to the abnormally long bones.) As CA also accurately describes the clinical signs in 'fawn calves' according to standard scientific disease nomenclature, for consistency and comparative purposes, the term CA will be adopted as the official name for this genetic defect in cattle.

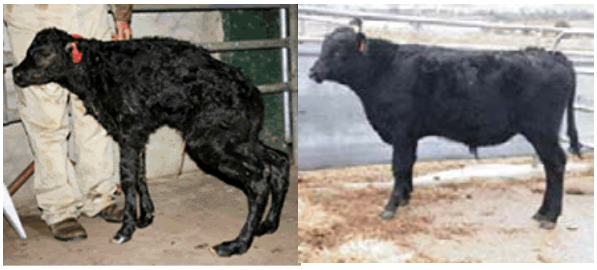

Figure 7. CA case at 1 and 260 days of age

Figure 8. CA at 6 years, same case as Fig. 7.

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