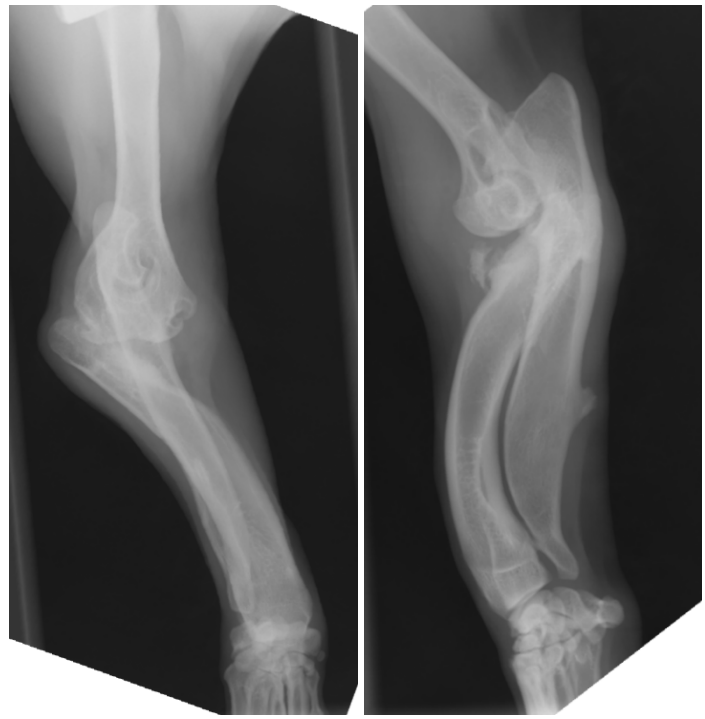
Chondrodysplasia in Bouviers

Radiographic findings in Bouviers with a radius curvus syndrome



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Abstract

Bouviers are a popular breed of dog in Holland. Since some time the breed association has noticed that in different litters pups were born with deformed forelimbs. The abnormalities seen in different degrees in the young Bouviers include: outwardly turned feet, inwardly turned elbows and short legs, some of the dogs show symptoms of lameness. The defect appears to be hereditary and seems to fit into the syndrome of chondrodysplasia.
Lameness in the front limbs is a common complaint for owners to seek out veterinary help. The causes can be categorized into congenital anomalies, developmental disorders, traumatic, idiopathic, infectious, nutritional, metabolic and neoplastic disorders. The first two are more common and can have a genetic basis.
28 Bouviers, both with and without the skeletal anomaly, have been clinically and radiographically examined. The age ranged from 6 weeks to 11 years. The most prominent abnormalities found, include (sub)luxation of the radial head and bowing of the proximal radius in a caudolateral direction, bowing of the ulna, a shallow trochlear notch, a widened olecranon fossa and (sub)luxation of the ulna at the level of the carpus and valgus deviation of the front paws. The abnormalities show a lot of similarity with CEL type 1 as reported by Kene et al (1982).

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Introduction

Bouviers are a popular breed of dog in Holland. Not many hereditary disorders circle in this population. Since some time however dog owners have noticed that in different litters pups were born with deformed forelimbs. The number of litters with pups showing this defect appears to have grown. For that reason the breed association (Nederlandse Bouvier Club) contacted the faculty of veterinary medicine in Utrecht. The faculty set up a couple of dates at which a number of Bouviers (both healthy and with the defect) were clinically and radiographically examined.

The abnormalities seen in different degrees in the young Bouviers include: outwardly turned feet, inwardly turned elbows and short legs (Hazewinkel 2008), see figure 1. Some of the dogs have problems in the elbow joint, causing them to show symptoms of lameness (Hazewinkel 2008). The defect appears to be hereditary and seems to fit into the syndrome of chondrodysplasia, a congenital lessening of cartilaginous growth (Hazewinkel 2008).

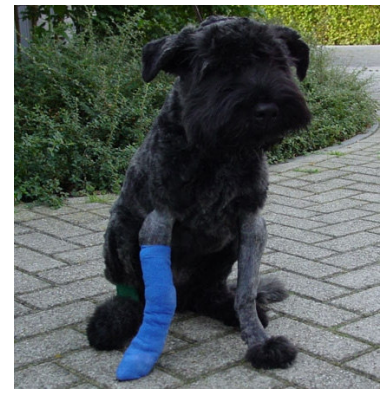

Figure 1 – Bouvier with abnormal front limbs

Lameness of the forelimb is a frequently seen problem in dogs under 1 year of age. Many causes are known (Cook 2001). The causes can be categorized into congenital anomalies, developmental disorders, traumatic, idiopathic, infectious, nutritional, metabolic and neoplastic disorders (Cook 2001). The first two can have a genetic basis and could be the cause of the lameness in the Bouviers. Infectious, nutritional, metabolic and neoplastic causes are infrequently seen (Cook 2001).

Much research has been dedicated to deformities of the limbs in dogs and a lot of articles have been published on the topic. The syndrome seen in the Bouviers does not seem to fit the most commonly described anomalies. There also seems little known about the defect in the Bouviers; no specific literature is available on growth disorders in the forelimb of Bouviers.

The goal of this article is to describe and inventory the abnormalities seen on the radiographs of the front legs of the Bouviers and to find clues for an etiology (and thus pointing out areas for further research).

I will start by describing the normal development and anatomy of the forelimb. After that I will recount what is written in the literature on growth disorders that could fit the abnormalities seen in the Bouviers. Next I will describe the defects seen on the radiographs, followed by a discussion of similarities and differences with the literature and the possible etiologic factors.

Normal anatomy and development of the forelimb

The forelimb is built up of the brachium, antebrachium, carpus and manus. The brachium or upper arm consists of the humerus. At the proximal end of the humerus there is a round caput that contacts the glenoidal cavity of the scapula (Dyce 2002). The tuberculum major and minus, separated by the intertubercular groove, border the caput (Dyce 2002). A spiral groove runs over the lateral side of the shaft (Dyce 2002). At the lateral side of the shaft the deltoid tuberositas is located and connected to the tuberculum major through a prominent ridge (Dyce

2002). On the medial side of the shaft the tuberositas teres major is located (Dyce 2002). The distal end of the humerus consists of an articular condyle, divided in a medial area (trochlea) for the ulna and a lateral area (capitulum) for the radius (Dyce 2002). The caudal part of the trochlear groove is continued proximally in a deep fossa olecranon, that offers space for the anconeal process of the ulna (Dyce 2002). Proximal of the articular part are a medial and a lateral epicondyle (Dyce 2002). The foramen supratrochlearis perforates the bottom of the fossa olecranon an opens in the hollow fossa radialis in the cranial part of the shaft (Dyce 2002).
The antebrachium or lower arm is formed from two bones: radius and ulna. In the standing animal the ulna is caudal from the radius in the proximal part of the antebrachium, but lateral in the distal part (Dyce 2002). The radius and ulna only make contact at the ends, making pronation and supination (up to 45°) possible (Dyce 2002). The radius is shaped like a rod, widening at the proximal end, where it articulates with the humerus, and at the caudal side with the ulna (Dyce 2002). The shaft is cranio-caudally compressed and slightly bowed in length (Dyce 2002). The distal end is somewhat enlarged and has a slight concave oval form, allowing for abduction, adduction and rotation in the carpus (Dyce 2002). Medial to the carpal joint the radius is lengthened and forms the medial styloid process (Dyce 2002). The ulna has a small shaft and is lengthened proximal to the elbow by the olecranon process (Dyce 2002). Distal to the olecranon on the cranial border is the anconeal process and further distally there is a facet for articulation with the radius (Dyce 2002). The shaft is equal in length to the radius and both bones are separated by a membrane crossing the interosseus space (Dyce 2002). The distal end has a facet for articulation and ends in the lateral styloid process (Dyce 2002).
The short carpal bones are located in two rows. The proximal row contains the radial, ulnar and accessory bone (Dyce 2002). The accessory bone extends caudal from the carpus (Dyce 2002). The distal row is numbered 1 through 4 (Dyce 2002). On the medial side of the carpus is a small sesamoid bone (Dyce 2002).
Most movements in the carpus occur at the level of the antebrachiocarpal joint, some in the intracarpal articulation and almost none in the carpometacarpal articulation (Dyce 2002).

The first signs of the developing limbs are paired projections growing from the ventrolateral surface of the embryo. In dogs these can be seen from the fourth week of gestation (McGeady 2006). The growing extremity consists of a mesenchymal core and an outer layer of ectoderm (Dyce 2002, McGeady 2006). The first indication of the skeleton consists of a thickening of the central mesoderm forming a core in the limb bud, that later develops into a series of cartilage models shaped like the future mature bones (Dyce 2002, McGeady 2006, Bingel 1977). In the next stage the cartilage is replaced by bone tissue. The ossification in a typical long bone takes place through two processes (Dyce 2002, McGeady 2006).
The first process is intramembranous ossification. The perichondrium around the middle of the shaft lays bone on the cartilage within a connective tissue membrane, this causes a tubular bone layer (a collar) to form around the centre of the shaft, that slowly expands to the ends (Dyce 2002, McGeady 2006). With the second process the cartilage in the centre of the shaft degenerates and the matrix is impregnated with calcium salts (Dyce 2002, McGeady 2006). Next connective tissue from the periosteum grows into the isle of dead cartilage, bringing with it several cell types with different functions responsible for the process of endochondral ossification (Dyce 2002, McGeady 2006). The balanced construction and destruction change the middle of the shaft into the primary or diaphysary centre of ossification (Dyce 2002, McGeady 2006). Later similar centres of ossifications are formed at the two extremities of the cartilage model: secondary or epiphysary centres of ossification (Dyce 2002, McGeady 2006). Bone growth is more complex then described above. The required size and shape can only be

obtained by remodelling the bone, a combination of bone deposition and resorption (Dyce 2002, McGeady 2006).
Eventually only two plates of the original cartilage remain: the epiphisiary or growth plates, situated between the primary and secondary centres of ossification. (Dyce 2002, McGeady 2006). Interstitial growth of the cartilage in these growth plates ensures the lengthening of the bone, as long as the growth speed equals the speed of endochondral ossification (Dyce 2002, McGeady 2006, Hazewinkel 1998). With the aging of the animal the replacement of cartilage by bone goes faster than the cartilaginous growth, causing the plate to thin and eventually disappear (Dyce 2002, McGeady 2006). After fusion between epiphysis and diaphysis further growth in length is impossible. Times of closure of the growth plates in different bones and of different growth plates in one bone can be very variable (see table 1, Dyce 2002, McGeady 2006). Appositional growth underneath the periosteum combined with intramedullar bone resorption increases the diameter of the bone and ensures a controlled thickening of the cortex (McGeady 2006, Hazewinkel 1998).

Table 1 Closure of the growth plates in dogs as visible on radiographs (Dyce 2002, Hazewinkel 1998)

Growth plate	Time of closure on radiographs
Radius	
proximal epiphysis	5-11 months
distal epiphysis	6-12 months
Ulna	
tuberculum olecranon	5-11 months
processus anconeus	3-5 months
distal epiphysis	6-12 months

The tissues for the synovial joints come from mesoderm that was left between the cartilage models of the bones (Dyce 2002, McGeady 2006). Spaces develop in the mesoderm, which coalesce to form the synovial cavity (Dyce 2002, McGeady 2006, Bingel 1977). The cavity is bordered by joint cartilage and the synovial membrane. The joint cartilage forms through late chondrification of the mesoderm on the edge of the cartilage models (Dyce 2002). The synovial membrane is a more direct transformation of the mesoderm at the border of the synovial cavity, while the fibrous part of the capsule and the peri-articular ligaments develop from more peripherally located mesoderm (Dyce 2002, McGeady 2006, Bingel 1977).

The skeleton of dogs is very immature at the time of birth; many of the secondary centres of ossification still need to be formed (Dyce 2002). The individual speed of development of the skeleton is dependent on many factors: hereditary, nutrional, hormonal (Dyce 2002, McGeady 2006). This combined action allows many possibilities for mistakes, making it unsurprising that skeletal anomalies occur frequently (Dyce 2002, McGeady 2006). Growth in length and development and maturation of secondary ossification centres are dependent on the breed of dog and can vary between individuals in one breed (Hazewinkel 1998).

The growth of paired bones is highly orchestrated so that both bones continue to form congruent joint surfaces at both ends (Hazewinkel 1998, Weigel 1987). This is especially remarkable in the radius and ulna: in the radius, 60 to 75% of bone growth occurs at its distal growth plate and 25-40% at its proximal growth plate, whereas the distal growth plate of the ulna accounts for approximately 80-90% of its growth in length and the proximal plate for the remaining 10-20% (Hazewinkel 1998, Gurevitch 1980, Johnson 1981, Weigel 1987). Because

of this difference the radius and ulna shift in relation to each other during the growth period, but the elbow joint surface remains congruent (Hazewinkel 1998, Weigel 1987). As the elbow joint is composed of three articulations: humero-ulnar, humero-radial and proximal radio-ulnar, displacement of one of the bones influences two of the component articulations (Kene 1982). The distal radius and ulna keep in line with the carpal bones as well to form a normal antebrachiocarpal joint (Hazewinkel 1998).
Not surprisingly many things can go wrong during the development of the antebrachium. Various causes result in different growth lengths of the antebrachium. In some cases this abnormality causes lameness and/or abnormal leg configuration.

Disorders of the forelimb in the growing dog

Disorders of the front leg are a frequent cause of concern for dog owners, especially in the young dog. Many disorders can cause lameness and deformity of the leg. These can be divided into seven categories: congenital, developmental, traumatic, idiopathic, infectious, nutritional/metabolic and neoplastic (Cook 2001). The Bouviers that were examined for this research showed the following clinical signs: outwardly turned feet, inwardly turned elbows and short legs. Symptoms sometimes referred to as the 'radius curvus' syndrome (Hazewinkel et al. 1998).
Disorders described in the literature that can cause these symptoms include: asynchronous growth of radius and ulna, retained cartilaginous cones and congenital elbow luxation or subluxation.

Asynchronous growth of the radius and ulna

Asynchronous growth between the radius and ulna occurs at a microscopic level in all dogs, but this is quickly compensated, so that macroscopically the involved joints stay congruent, the bones are equal in length and aligned properly and the function is not impaired (Cook 2001, Hazewinkel 1998). Asynchronous growth pathologically causes an angular limb deformity and joint incongruity and may be responsible for ununited anconeal process (UAP) and fragmented medial coronoid process (FMCP) (Cook 2001, Hazewinkel 1998). These problems usually are noted before the dog is 12-18 months old (Cook 2001). Clinically asynchronous growth can be seen as joint incongruity and abnormal bone modelling (Cook 2001). The joint incongruity leads to subluxation and abnormal biomechanics and secondary osteoarthritis can be seen (Cook 2001). The abnormal bone modelling leads to radioulnar bowing and rotation, which can occur without joint incongruity (as seen in the chondrodystrophic breeds) (Cook 2001).

Asynchronous growth of the radius and ulna in the dog can have several causes: hereditary, trauma, imbalanced nutrition, hypertrophic osteodystrophy, septic physitis and idiopathic (Hazewinkel 1998, Cook 2001).

The hereditary form of asynchronous growth of radius and ulna is frequently seen in the chondrodystrophic breeds, but can sometimes be seen in pups from nonchondrodystrophic breeds with symptoms of chondrodysplasia (Hazewinkel 1998, Cook 2001, Gurevitch 1980). These dogs display a decreased growth in length of all long bones, however this is most pronounced in the ulna (Hazewinkel 1998). On ML-radiographs the distal ulnar growth plate has an oblique shape rather then the typical V-shape seen in dogs from other breeds, visible only in dogs younger than 6 months (Hazewinkel 1998). Furthermore the radius is extended proximally beyond the joint level of the ulna and pushes against the humerus, that in turn

pushes against the anconeal process (Hazewinkel 1998). The result of this elbow incongruity can be an ununited anconeal process (dogs under the age of 6 months) or, in older dogs, a syndrome called distractio cubiti (Hazewinkel 1998, Demko 2005). The chondrodysplasia can affect individuals or entire litters (Hazewinkel 1998).

Trauma is the most commonly seen cause of asynchronous growth between radius and ulna (Cook 2001, Gurevitch 1980). The asynchronous growth can result from injury to any of the physes of the radius and ulna, although changes as a result of injury to the proximal ulnar physis are rare (Cook 2001). Most commonly trauma to the distal ulnar growth plate in the form of a Salter Harris type V injury, can cause a temporary or permanent disruption of the growth in length of the ulna (Hazewinkel 1998, Cook 2001, Gurevitch 1980, Johnson 1981). This can be the result of damage to the germinal cells or to the vascular supply of the growth plate (Hazewinkel 1998, Cook 2001). This damage may initially be invisible clinically and/or radiographically (Cook 2001). However, if the growth plates of the radius are still intact, the resultant growth discrepancy may cause the radius curvus syndrome (Hazewinkel 1998). This syndrome can include the following findings (Hazewinkel 1998):

- the ulnar styloid process is proximal to the accessory bone on mediolateral radiographs
- the lateral coronoid process is distal to the joint surface of the radial head
- the radius is curved on mediolateral radiographs, with a palmar cortex thicker and a dorsal cortex thinner than normal
- in more severe cases a cranio-caudal and valgus curving of the radius is present
- a valgus deformity of the distal extremity
- abnormal development of the antebrachiocarpal joint
- in more severe cases: ununited anconeal process or distractio cubiti

Imbalanced nutrition can cause deformity of the antebrachium in the growing dog. Three causes are clinically significant: excessive calcium intake, excessive nutrient intake and vitamin D deficiency (Hazewinkel 1998). Thanks to commercial, balanced pet foods however skeletal abnormalities secondary to nutritional abnormalities are rare (Cook 2001, Hazewinkel 1998).
A high calcium intake impaires the maturation of cartilaginous cells, which leads to a disturbance of the endochondral ossification (Hazewinkel 1998). Direct or indirect effects of calcium on the cartilage cells or a disturbance of the vascular supply may be the cause of the disturbed endochondral ossification (Hazewinkel 1998, Cook 2001). On radiographs the disturbed ossification is seen as a retained cartilage cone in the metaphyseal area and a flattening or indentation of the distal ulnar metaphysis (Hazewinkel 1998). Studies in Great Danes showed that a clinical ‘radius curvus’ syndrome only develops when the cones exceed 20 to 25 mm in length (due to decreased growth in length of the ulna), smaller cones will spontaneously disappear at 6 months of age (Hazewinkel 1998).
Excessive nutrient intake occurs when the pup is fed an adult diet that is relatively high in calcium or when a food supplement of calcium is fed (Hazewinkel 1998). As the growing puppy has higher caloric requirements then the adult dog, the pup will eat extra food to obtain these calories and, when fed an adult diet, with this extra food the pup also takes in additional calcium (Hazewinkel 1998). This excessive nutrient intake and thus the higher calcium intake will cause abnormalities as described above, in addition osteochondrosis of joint cartilage can occur (Hazewinkel 1998).
A vitamin D deficiency also results in disturbed endochondral ossification (Hazewinkel 1998). The hypovitaminosis D leads to a lack of matrix mineralization and thus postpones the cell death of the cartilage cells (Hazewinkel 1998). Radiographically the growth plates will be

widened and a varus or valgus deformation (due to bowing of the weakened long bones) can be seen (Hazewinkel 1998). Dogs are unable to synthesize vitamin D, the main cause of hypovitaminosis D therefore is a diet lacking in vitamin D, for example a diet of only lean meat (Hazewinkel 1998, Cook 2001). Clinically signs of rickets develop including soft tissue swelling around the metaphysis of the distal radius (Hazewinkel 1998). The abnormalities resolve after feeding a commercial diet (Hazewinkel 1998).

Hypertrophic osteodystrophy can lead to valgus deviation of the front paws (Hazewinkel 1998) In this disease the growth in length of all long bones is disturbed, however the growth plates with the largest growth potential are most commonly affected (Hazewinkel 1998). The cause is unknown, but an association with distemper virus is suspected (Hazewinkel 1998). The most prominent clinical signs include depression and extreme pain on palpation of the swollen metaphyseal areas (Hazewinkel 1998). On radiographs a radiolucent line parallel to and 2-3mm from the growth plates is pathognomonic in the subacute phase (Hazewinkel 1998).

Retained cartilaginous cone

Retained cartilaginous cones are a result of delayed enchochondral ossification in (part of) the distal ulnar metaphysis (Cook 2001, Hazewinkel 1998, Johnson 1981). They are mainly seen in dogs from large and giant breeds between 4 and 8 months of age (Cook 2001, Thrall 2002). As a result of abnormal ossification, the hypertrophied chondrocytes and the extracellular matrix are not transformed into bone and the cartilage is retained in the growing ulna (Cook 2001, Hazewinkel 1998, Thrall 2002, Johnson 1981). This retained cartilage is visible on radiographs as a cone-shaped radiolucent area extending from the distal ulnar physis (Cook 2001, Hazewinkel 1998, Thrall 2002, Johnson 1981). A narrow zone of sclerosis may surround the radiolucent area (Thrall 2002). The etiology is unknown, but a multifactorial process is suspected (Cook 2001). The delayed ossification can result in a slowed growth in length of the ulna, and thus joint and bone abnormalities associated with asynchronous growth of radius and ulna (Cook 2001, Johnson 1981). Clinical signs include a weightbearing lameness, distal ulnar pain and swelling and angular and/or rotational deformities (Cook 2001, Thrall 2002). Most dogs are only minimally affected and do not require treatment (Cook 2001, Hazewinkel 1998, Thrall 2002). In more severe cases, however, surgical intervention is necessary (Cook 2001).

Congenital elbow luxation

Congenital elbow luxations and subluxations (CEL) are described in the literature as a separate group of abnormalities apart from elbow dislocation and other elbow deformities, such as patella cubiti and defects related to disturbance of the growth plate (Bingel 1977, Milton 1979, Pass 1971). CEL is reported in the literature to be responsible for 17-20% of cases of non-traumatic (Pass 1971, Stevens 1974, Campbell 1969, Milton 1987). CEL primarily affects small breeds of dogs, but it has also been reported in larger breeds (Gurevitch 1980, Kene 1982, Milton 1979, Bingel 1977, Cook 2001). The lesion can be seen either unilateral or bilateral (Kene 1982, Milton 1979), although bilateral involvement appears to be more common (Milton 1979, Cook 2001). Laxity of the elbow joint (CEL type 3) can be seen in pups with multiple congenital defects, such as polyarthrodysplasia or ectrodactyly (Milton 1979 & 1987, Bingel 1977, Cook 2001).

The disease is considered hereditary (Cook 2001). The etiology is thought to involve agenesis or hypoplasia of the medial collateral ligament, abnormal development of the humeral trochlea and anconeal process, or both (Cook 2001, Milton 1987). Affected dogs should not be bred (Cook 2001).
The term CEL is used to describe conditions varying from mild lateral subluxation of the radial head with minimal lameness to severe deformity of the elbow, bowing of the leg and an inability to extend the elbow during attempted weight bearing (Gurevitch 1980, Milton 1979 & 1987, Bingel 1977, Cook 2001). The symptoms are varying degrees of pain, lameness and dysfunction and can be noticed by the owner when the pup is only 3-6 weeks old (Cook 2001, Milton 1979, Bingel 1977, Kene 1982). On physical examination the puppies have a varus deviation of the antebrachium (Cook 2001). The diagnosis can easily be made based on signalment, history, physical examination and radiographic findings (Cook 2001, Kene 1982).

Radiographically CEL is categorized into three types (Kene 1982, Cook 2001, Peirone 2004, Milton 1987):

- type I is described as a humeroradial luxation, where the proximal radius is displaced caudolaterally and does not articulate with the humeral capitulum, the ulna appears in a relatively normal position
- type II involves luxation of the humeroulnar articulation: the ulna is displaced in a laterally rotated manner, while the proximal radius had a normal anatomic relationship to the humerus
- type III is described as dislocation or joint laxity and affects both humeroradial and humeroulnar articulations, with the radius and ulna displaced laterally to the humeral condyle and rotated approximately 90° to the sagittal plane of the humerus

The treatment depends on the age of the animal, the severity of the luxation and clinical signs, the degree of secondary changes present and the intended use of the dog. It is best to treat dogs as early as possible to potentiate successful reduction and minimize the secondary growth abnormalities and osteoarthritis that often occur (Cook 2001, Peirone 2004). Conservative management is not effective (Cook 2001). The treatment involves open or closed reduction techniques, sometimes combined with ostectomy and internal and external fixations (Cook 2001, Milton 1987). The prognosis is guarded to poor (Cook 2001).

Materials and methods

The study population includes 28 Bouviers, both with and without the skeletal anomaly. The age ranged from 6 weeks to 11 years. Both littermates and animals unrelated to each other were examined. A thorough clinical examination followed by a radiographic examination of the right front leg was performed in all 28 dogs.
Radiographic examination of the right front legs included a mediolateral and a craniocaudaal projection.
For the mediolateral views the dogs were placed in lateral recumbency with the elbow being examined directly on the cassette in an extended position. The craniocaudaal radiographs were taken with the dog in ventral recumbency, with the elbow being examined lying directly on the cassette in an extended position.
The seven youngest pups (all from the same litter) were examined at 6 weeks at which point the skeleton was not enough developed to draw any conclusions, therefore they were re-examined at 2 months and again at 4 months to follow their development. Of the other animals radiographs were taken once.

Results

All Bouviers showed similar changes of the antebrachium. However the degree of abnormality differed between individuals. The following description contains all defects that have been noticed in the dogs. For further details on abnormalities found in affected, individual dogs see table 2 and figures 2-5.

Of the 28 dogs examined, 16 animals showed bony abnormalities. Of the affected animals 6 were males and 7 were females, the sex of the other 3 animals was not known. The age of the affected dogs at examination ranged from 6 weeks to almost 5½ years (see table 2 for ages of individual dogs).

The elbow joint was incongruent, the joint surface of the ulna was more distal than that of the radius. The radial head was (sub)luxated and bowed. The amount of bowing of the radius varied, ranging from an almost normal contact with the humerus to being completely luxated. The ulna still made reasonably good contact with the humerus. Some dogs had a superficial trochlear notch. The joint space had an uneven thickness. The humerus had a widened fossa olecranon in most dogs.
The proximal radius was bowed in a caudolateral direction from approximately halfway the proximal metaphysis. On the ML radiographs the proximal radius was bowed caudally and more or less displaced, whereby it partially overlapped the proximal diaphysis of the ulna. In the CrCd projections the proximal radius was bowed laterally. In most dogs the radial epiphysis appeared to be normal to enlarged in size. In most dogs the radial head still made contact with the humerus, but in some it no longer articulated with the lateral condyle of the humerus.
The ulna was bowed in some dogs, while in others it pertained an almost normal outline. On the ML projections the proximal ulna was curved slightly cranial in most dogs, but the humeroulnar articulation appeared little affected. The distal part of the ulna was curved somewhat caudal on the ML views. On the CrCd radiographs the ulna appeared straight. (In some dogs the anconeal process seemed relatively small in size (compared to other dogs of the same size and age), while in the other dogs it appeared to be normal in size. Several dogs appeared to have a comparatively small medial coronoid process), although the borders of the medial coronoid process could not be clearly identified in all dogs.
The cortex of both radius and ulna showed an uneven thickening. The radial cortex was thickened at the concave part of the shaft (caudally when viewed on the ML radiographs). The ulnar cortex was thickened at the proximal part of the shaft (caudally when viewed on the ML radiographs). Overall both the radius and the ulna were shortened and thickened when compared to normal dogs of the same breed and age.

The carpus showed a valgus deviation. The radius was dislocated cranially over the carpal bones, but still made reasonably good contact with the carpal bones. The distal ulna was curved caudally when viewed on the ML radiographs and did not contact the carpal bones. The ulnar styloid process was small and irregularly shaped in most dogs. The antebrachiocarpal joint space was uneven.

Osteoarthritic changes were visible on the humeral epicondyles, olecranon, radial head, ulnar shaft and coronoid process in all dogs over 9 months of age. In some dogs a spur like type of new bone formation was seen in the caudal aspect of the middistal ulna, likely representing an enthesiophyte.

A litter of seven young pups was examined at 6 weeks, 2 months and 4 months of age. When examined at 6 weeks the skeleton was not enough developed to make out any abnormalities. However when re-examined at 2 months the pups had a small defect in the distal growth plate of the ulna. The growth plate did not show the normal V-shape, but seemed to be slightly rounded and widened at the metaphyseal side. Two of the seven pups had a more severe flattening of the distal ulnar growth plate on the ML radiographs and on the CrCd views had an irregular metaphyseal end of the distal ulnar growth plate with a large central defect looking somewhat like a retained cartilage cone.
At 4 months of age all but one of the pups showed a normal V-shape of the distal ulnar growth plate. One pup still showed a flattening of the metaphyseal part of the distal ulnar growth plate. The five pups with only slightly rounded growth plates all showed a normal development of the front legs. However the two pups with the more severely affected growth plates had a curved radius. The proximal radius was bowed in a caudolateral direction from approximately halfwaythe proximal metaphysis. On the ML radiographs the proximal radius was bowed caudally and slightly displaced (partially overlapping the proximal diaphysis of the ulna). In the CrCd projections the proximal radius was bowed laterally. In one pup the distal ulna appeared to bow slightly caudally when viewed on the ML radiograph.

In conclusion the most prominent features of the condition of the Bouviers are:

- (sub)luxation of the radial head and bowing of the proximal radius in a caudolateral direction
- slight bowing of the ulna (in a cranial direction proximally and a caudal direction distally)
- shallow trochlear notch
- small anconeal process and small coronoid process
- spur like new bone formation caudal aspect middistal ulna
- shortened and thickened ulna and radius
- widened olecranon fossa
- (sub)luxation of the ulna at the level of the carpus and valgus deviation of the front paws

Table 2 – Data on individual dogs examined

Name	Age	Sexe M/F/U	(Sub)luxation radius	Valgus	Bowing ulna	Widened fossa olecranon	Shallow trochlear notch	Small anconeal process	Small medial coronoid process	Growth plates closed	Osteoarthrosis
Pup 2	2mnd	F	SL	A	P	A	A	A	A	A	N
Pup 4	2mnd	F	SL	A	A	A	A	A	A	A	A
Boomer	9mnd	M	SL	P	P	P	P	A	A	A	A
Borus	9mnd	M	SL	A	P	P	P	A	A	A	A
Kateman	10mnd	F	SL	A	P	P	A	A	A	P	A
Luna	9mnd	F	SL	A	P	P	P	A	P	A	A
Chanel	10mnd	F	L	P	P	P	P	A	P	P	A
Derrick	10mnd	M	L	P	P	P	P	P	P	P	A
Kyra	10mnd	F	L	A	P	P	P	P	P	P	A
Snoop	10mnd	M	L	P	P	P	P	A	A	P	A
Beer Megan	1jr, 4mnd	M	L	P	P	P	A	A	A	P	P
Bello	1jr, 4mnd	M	SL	A	P	P	P	A	P	P	A
Macclaven	1jr, 4mnd	F	L	P	P	P	P	P	P	P	P
Bonita	5jr, 4mnd	U	SL	P	P	P	P	A	P	P	P
Zowie	U	U	SL	A	P	P	P	A	P	A	A
Beer	U	U	L	A	P	P	P	A	A	P	P

A = absent; F = female; L = luxation; M= male; P = present; SL = subluxation; U = unknown.

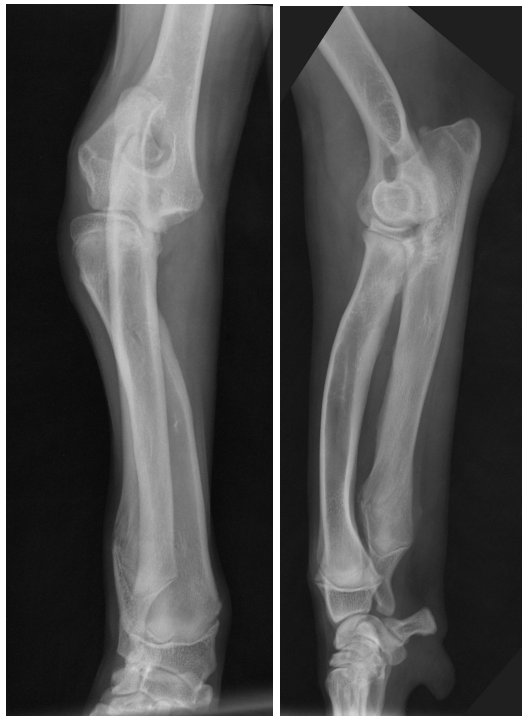

Figure 2 – Boomer (9 months of age)

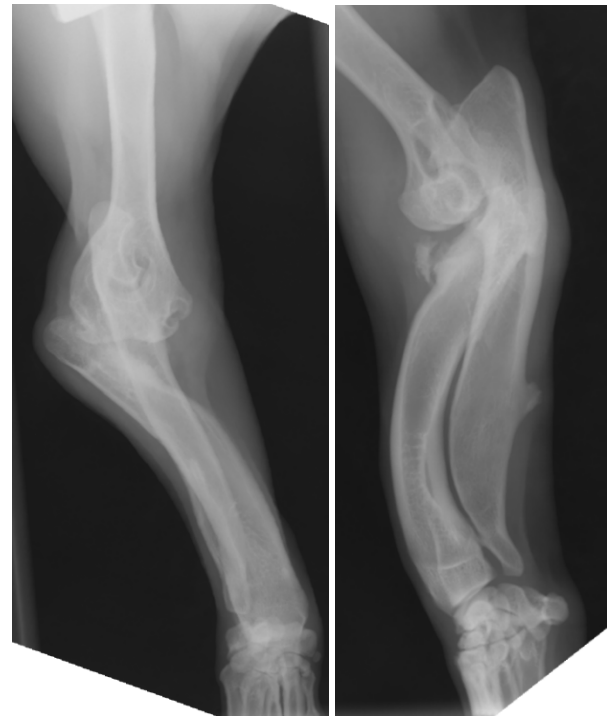

Figure 3 – Beer Megan (1 year, 4 months of age)

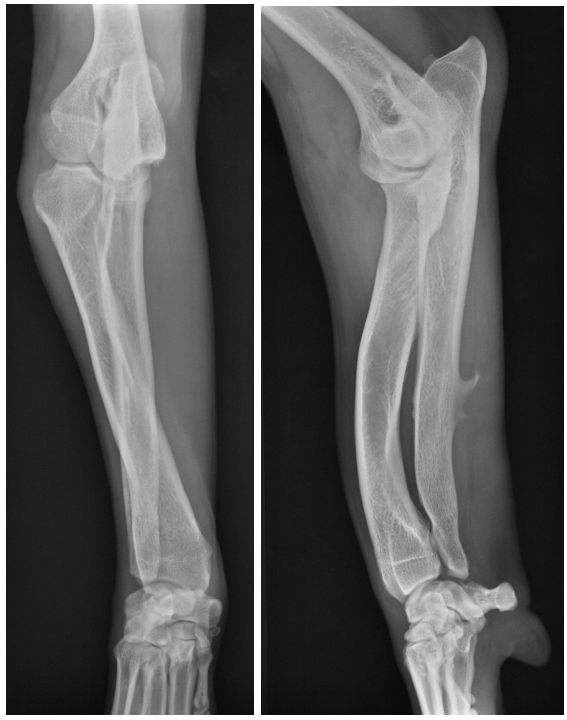

Figure 4 – Bonita (5 years, 4 months of age)

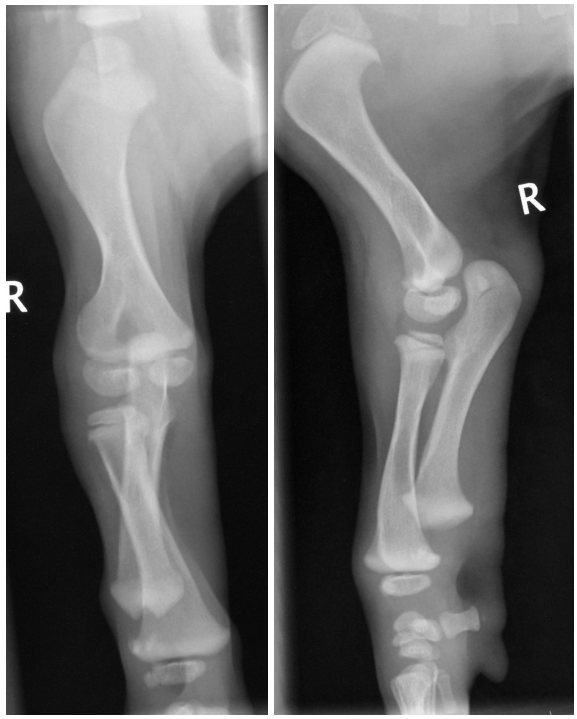

Figure 5a – Pup 2 (2 months of age)

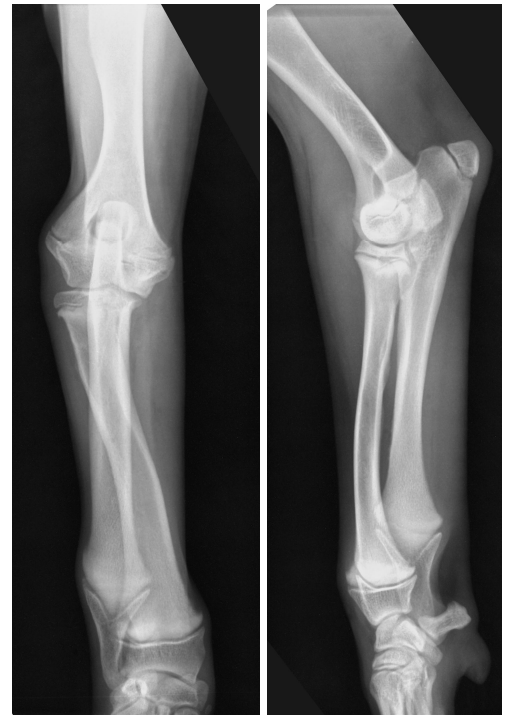

Figure 5b – Pup 2 (4 months of age)

Discussion & conclusion

The abnormalities seen with the Bouviers show a lot of similarities with CEL type 1 as described by Kene et al (1982). Kene et al (1982) described a caudolateral dislocation of the proximal radius as the most prominent feature of CEL type 1. Other abnormalities that were found included an underdeveloped proximal radial epiphysis, a convex, and sometimes enlarged, radial metaphysis, a narrow radial neck, cranial bowing of the proximal ulna, a small or absent anconeal proces, a small medial coronoid process, a superficial trochlear notch, an abnormally large olecranon fossa and a disrupted radio-ulnar articulation (Kene et al 1982, Milton 1987). In this study most abnormalities found were similar to those described by Kene et al (1982), however some differences were noted. Neither the radial epiphysis nor the radial neck appeared to be underdeveloped, the radial ephiphysis appeared normal to enlarged in size in all dogs. Furthermore the anconeal process and the coronoid process did not appear small or underdeveloped in all dogs, in fact they appeared normal in size in the majority of the Bouviers. The proximal radio-ulnar articulation did not appear to be disrupted. Kene et al 1982) did not report abnormalities in the cortex, however in this study a thickening of the cortex of the radius and ulna were seen. This thickening of the cortex probably provides a mechanism of compensation for the abnormal forces acting on the bones due to the abnormal position.

The abnormalities seen in the carpus have also been described in the literature in combination with CEL, but can also be found as part of the problems associated with asynchronous growth of the radius and ulna and a retained cartilaginous cone (Kene 1982, Lau 1977). The cranial dislocation of the distal radius seems to be responsible for the valgus deviation of the paws in some dogs. The dogs with the most deviated stance also showed the most dislocation of the radius.

In the young growing animal a defect in the distal ulnar growth plate seems to precede the development of the bony abnormalities. The appearance of the growth plate at 2 months of age shows similarities with the findings of a retained cartilaginous cone (Hazewinkel, 1998). At a later age however no indications of a cartilaginous cone were found in the metaphysis of the ulna. Kene et al (1982) did not find any abnormalities of the distal growth plates of radius and ulna, even though the dogs they examined were 1-5½ months.

Kene et al (1982) did not find any reactive bone changes, however the age of the examined dogs ranged from 1 to 5½ months. In this study osteoarthritic changes were found in all dogs over 9 months of age. The osteoarthrosis most likely developed secondary to the bony changes. It could be that the dogs in the study of Kene et al (1982) were to young to have developed signs of osteoarthrosis. Other authors did report reactive bone changes in combination with CEL (Stevens 1974, Milton 1979).

It is suggested that a shortened ulna could be responsible for a limited growth of the radius, creating a ‘bowstring’ effect (Gurevitch 1980, Carrig 1975). Other deformities resulting from this ulnar bowstring effect are: valgus deviation of the paw, shortening of the limb, outward rotation of the carpus and metacarpus, increased separation between the radial and ulnar shafts, caudal subluxation of the carpus, different forms of elbow subluxation and secondary joint injuries of carpus and elbow (Gurevitch 1980, Carrig 1975).

In the literature it is reported for type 1 CEL that medium and large size breeds of dogs 3-4 months of age are mostly affected (Peirone 2004). The Bouviers in this study can be

categorized as a large breed of dog and support this statement. The affected leg shows a mild deformity and dysfunction, on physical examination the luxated radial head can be palpated and the dog doesn't show pain on manipulation (Peirone 2004).

Gurevitch (1980) suggests that the most common cause of lateral luxation or subluxation of the radial head in dogs is premature closure of the distal ulnar growth plate, even though distal luxation or subluxation of the ulna is a more common consequence (Gurevitch 1980). Gurevitch (1980) reported 43 cases of radius curvus as a result of abnormalities in the distal physis of the ulna or parts of the distolateral physis of the radius, of which only 7 showed (sub)luxation of the radial head as the primary problem (Gurevitch 1980). All 7 dogs had achondroplastic features (Gurevitch 1980).
The premature closure of the distal ulnar physis usually is the result of trauma, but can also be caused by radiation, nutritional imbalances and congenital factors, such as retained hypertrophied endochondral cartilage in the ulnar metaphysis (Gurevitch 1980, Lau 1977). In this study the distal ulnar growth plate appeared open in dogs up to 9 months of age, it was closed in all older dogs. In the literature it is reported that closure of the distal ulnar growth plates occurs between 6 and 12 months of age (Dyce). It is therefore difficult to conclude whether closure around 9 months is normal or abnormal in these dogs. Unfortunately no normal Bouviers around 9 or 10 months of age were included in this study so a comparison could not be made. As described above however there were some findings suggesting the possibility of a retained cartilaginous cone that could be responsible for a premature closure of the distal ulnar growth plate (Gurevitch 1980, Hazewinkel 1998, Cook 2001)
Gurevitch reported that congenital (sub)luxations of the radial head have a different pathogenesis then those secondary to premature closure of the growth plate (Gurevitch 1980). In Skye Terriers hereditary subluxation of the radial head secondary to premature closure of the distal ulnar growth plate has been described by Lau (1977). The findings in this study show similarities with those reported by Lau (1977). However the dogs examined by Lau (1977) were 3-5 months of age and already had a closed distal ulnar physis. Lau (1977) reported that the premature closure of the distal ulnar growth plate is a recessively inherited trait in Skye Terriers.

In the literature an etiology for CEL is proposed concerning an underdevelopment of the intra-articular ligaments (Bingel 1977, Gurevitch 1980, Milton 1979 & 1987). Bingel suggests that the pathogenesis of CEL is related to an embryonic failure at the formation of the intra-articular ligaments, mainly aplasia or hypoplasia of the medial collateral and annular ligaments (Bingel 1977, Gurevitch 1980). The weakened medial collateral ligaments would allow external rotation of the proximal ulna and subluxation of the radial head, the ulna or both (Bingel 1977, Gurevitch 1980). The hypoplastic annular ligament would allow subluxation of the radial head (Bingel 1977, Gurevitch 1980). The abnormal development of the joint would be a result of the abnormal dislocations of the bones of the elbow (Bingel 1977). However Milton et al (1979 & 1987) reported surgical observations and anatomical dissections that did not agree with the theory that hypoplasia of the medial collateral ligament is the primary etiological factor for CEL (Milton 1979 & 1987). They reported that the medial collateral ligament seemed to give little support to the ulna, especially regarding the rotational stability (Milton 1979 & 1987). Milton et al (1979 & 1987) suggest no clear cause for CEL can be appointed from the studies (Milton 1979 & 1987).

In this study it was not possible to examine the intra-articular ligaments. Further research should be performed to determine whether abnormalities of the intra-articular ligaments could be responsible for the abnormalities seen in the Bouviers or in other cases of CEL.

Hypoplasia or aplasia of the anconeal and medial coronoid processes have been described as some of the abnormalities seen in CEL (Kene 1982, Bingel 1977, Gurevitch 1980). As has a superficial trochlear notch, though less frequently encountered (Kene 1982). It is unclear however whether these findings have significance as a primary or secondary defect (Bingel 1977, Gurevitch 1980, Kene 1982)
Kene et al (1982) were unsure whether these abnormalities could be part of the CEL complex since they only reported examinations of dogs under 5½ months of age. In this study however these abnormalities were also found in some of the dogs over 1 year of age, suggesting they are part of the complex of abnormalities found with CEL. The superficial trochlear notch for example could be a consequence of the nondevelopment of the anconeal and medial coronoid processes (Kene 1982).

It is unclear in the literature whether CEL is a hereditary condition. A possible hereditary basis was suggested for the following reasons: (1) a high incidence of bilateral involvement; (2) appearance of CEL in more than one pup from a litter; (3) the embryonic stage at which the defect started; and (4) frequency of the condition in animals with multiple soft tissue and skeletal deformities (Bingel 1977, Milton 1979). Milton reported some findings that counteract this theory: no littermates of the pup from a larger litter were affected and one affected dog fathered a normal litter (Milton 1979). Further studies need to be performed to clarify the possible heritability of CEL (Milton 1979, Bingel 1977).
Apart from the discussion whether all types of CEL are a hereditary condition it has been questioned if CEL type 1 should be classified as hereditary (Milton 1979, Peirone 2004). The reason for this is that (sub)luxation of the radial head can be secondary to trauma or asynchronous growth between radius and ulna (Peirone 2004, Milton 1979, Gurevitch 1980). Milton et al (1979) also questioned whether the luxation or dislocation of the radial head should be considered congenital, since the condition can remain unnoticed for months or years and can also result from asynchronous growth between radius and ulna due to acquired disturbances of the growth plate (Milton 1979).
In this study several littermates from three different litters were affected. And in one litter of 6 pups two pups showed the abnormalities, while the other four showed a normal development of the antebrachium. This could suggest a possible hereditary basis, but as stated earlier further research is necessary to clarify this point.
The separation between congenital and developmental abnormalities is a very thin line. A congenital problem is present at birth and can be noticed in the youngest patients, while a developmental disorder occurs during the growth phases of the patient (Cook 2001). Both congenital and developmental problems can have a genetic basis. It is unclear whether the disorder affecting the Bouviers is congenital or developmental. The findings in this study suggest that the disorder is developmental, since it was not noticed until the pups were 2 months or older. However a congenital basis for the disease could already be in place, even if it is not yet visible on radiographs. Further research should make this clear.

In the literature it is reported that CEL can be seen both in male and female dogs, but male dogs seem to be overrepresented (Cook 2001, Milton 1979 & 1987). In this study however male and female dogs appear to be equally affected (6 males, 7 females).

In conclusion the abnormalities seen in the Bouviers seem to be part of a combined complex of abnormalities not previously described. Nevertheless a lot of similiarities can be found with CEL type 1 as reported by Kene et al (1982). Areas for further research should include the distal ulnar growth plate and the intra-articular ligaments to determine whether a retained

cartilaginous cone or abnormal ligaments could be responsible for the abnormalities seen in the Bouviers. Further research is needed to clarify these points.

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