

Information for Breeders: Hermaphrodites (XX sex reversal)

An Inherited Disorder of the Reproductive Tract

From: Vicki N. Meyers-Wallen, VMD, PhD, Diplomate of the ACT

XX sex reversal (XXSR) is an inherited disorder that causes infertility and sterility in some breeds of purebred dogs. At present, this inherited disorder is documented in 28 breeds, but dogs of other breeds are currently being examined.

TABLE 1: Canine breeds in which XX DSD has been identified. [¹Meyers-Wallen et al., 1995a; ²Meyers-Wallen et al., 1999; ³Meyers-Wallen, unpublished; ⁴Hubler et al., 1999; ⁵Williams et al., 1997; ⁶Poth et al., 2010; ⁷Campos et al., 2011; ⁸Switonski et al., 2004; ⁹Meyers-Wallen et al., 1995b; ¹⁰Kuiper et al., 2005; ¹¹Williamson, 1979; ¹²DeLorenzi et al., 2008; ¹³Melniczek et al., 1999; ¹⁴Buijtelts et al., 2009; ¹⁵Stewart et al., 1972; ¹⁶Rota et al., 2010]

American cocker spaniel ^{1,2}	German shorthaired pointer ⁹
Afghan hound ²	Golden retriever ³
American pit bull terrier ²	Jack Russell terrier ¹⁰
American Staffordshire terrier ³	Kerry blue terrier ¹¹
Australian shepherd ³	Mixed breed ¹²
Basset hound ⁴	Norwegian elkhound ¹³
Beagle ⁵	Podenco dog ¹⁴
Border collie ²	Pug ^{2,15,16}
Brussels griffon ^{3,6}	Soft coated wheaten terrier ²
Doberman pinscher ²	Tibetan terrier ³
English cocker spaniel ²	Vizsla ²
French bulldog ^{3,7}	Walker hound ²
German pinscher ³	Weimaraner ²
German shepherd dog ⁸	Wheaten terrier ³

Affected dogs have female chromosomes (karyotype 78,XX), although dogs with male sex chromosomes (78,XY) can be carriers of XXSR. Normal dogs with female sex chromosomes develop only ovaries. Dogs affected with XXSR usually develop both ovarian and testis tissue (ovotestes). On rare occasions, affected dogs develop only testis tissue (bilateral testes). As a result, the rest of the reproductive tract develops abnormally. This leads to infertility and sterility.

Externally

[*\(See our website for more pictures\)*](#)

Scroll down to **"2. SRY-negative XX sex reversal (XXSR)"**

Affected dogs can have any of the following signs:

A vulva that is shaped like a prepuce.

An enlarged clitoris (containing a bone). This structure often protrudes from the vulva in affected dogs, as in the picture below.



A longer distance between the anus and the vulva than is normally seen in females (the vulva may be located very near the belly), or a small prepuce.

If they have a penis-like structure they have hypospadias (the urethra does not open at the tip of the penis, but opens farther caudally).

Internally – Affected dogs usually have:

1. Bilateral ovotestes or testes
2. One ovotestis and one ovary
3. Rarely, two testes (these dogs look more like a male externally)

Most often, affected dogs cannot be differentiated from normal females during a spay/neuter operation even if the veterinarian is an expert in XXSR because testicular tissue is often in the center of the gonad and not at the surface. Carriers look like normal females or normal males. Correct diagnosis depends upon microscopic examination of the gonads by a pathologist who is familiar with XXSR. When this is done, either one ovary and an ovotestis, two ovotestes, or two testes are found.

When only testes are found, they are usually in the abdomen. Externally, these dogs look more like males than females, but if you look more closely they are not normal males. All affected dogs have a complete uterus. Many have an epididymis adjacent to the ovotestis or testis.

Also, all affected dogs have female sex chromosomes, as determined by chromosome analysis (karyotype 78,XX) and have no Y chromosome.

Polymerase Chain Reaction (PCR) tests are performed to determine the presence or absence of canine Sry in DNA from affected dogs. The Sry

gene is normally located only on the Y chromosome. Normal females and affected dogs do not have Sry in their DNA. Normal males have Sry.

At present, correct diagnosis of affected dogs is difficult (karyotype, DNA analysis, gonadal histology), and there is no practical method to identify carriers.

We hope to increase breeders' awareness of XXSR by describing the common findings in affected dogs. We need to know if this problem has been recognized in your breeds and how many dogs have been affected.

We are working to identify the gene mutation that causes this problem so that we can develop a DNA test. A DNA test will allow us to clearly identify both carrier and affected dogs, providing a practical method to improve breeding performance in these breeds.

You can help by contacting Dr. Vicki Meyers-Wallen if you think you have an affected dog. All information will be kept in the strictest confidence. If you wish, we can assist you and your veterinarian in diagnosis and genetic counseling.

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