



# Developmental Kidney Diseases

(Diseases That Are Inherited or Occur during Development of the Kidney)

## Basics

### OVERVIEW

- Congenital (existing at birth) and developmental kidney diseases are a group of diseases in which the kidney may be abnormal in appearance or may be abnormal in its ability to function normally or both. These diseases result from inherited or genetic problems or disease processes that affect the development and growth of the kidney before or shortly after birth. Examples of congenital and developmental kidney diseases include the following:
  - ♦ Failure of kidney formation (known as “renal agenesis”)—complete absence of one or both kidneys; one or both kidneys failed to form
  - ♦ Abnormal kidney development (known as “renal dysplasia”)—disorganized or abnormal kidney tissue development
  - ♦ Displacement of one or both kidneys (known as “renal ectopia”)—one or both kidneys are located in an abnormal position in the body at birth (congenital); ectopic kidneys may be fused together
  - ♦ Glomerulopathy—a group of small blood vessels in the functional unit of the kidney is the glomerulus; “glomerulopathy” is the broad name for any type of glomerular disease
  - ♦ Kidney disease involving the tubules and tissue spaces (known as “tubulointerstitial nephropathy”)—a non-inflammatory disorder of kidney tubules and tissue spaces (known as “interstitium”)
  - ♦ Polycystic kidney disease—characterized by formation of multiple, variable-sized cysts throughout the kidney tissue
  - ♦ Dilation of small blood vessels in the kidney (known as “renal telangiectasia”)—characterized by multiple blood-vessel malformations involving the kidneys and other organs
  - ♦ Amyloidosis of the kidney—(amyloidosis is a group of conditions of differing cause in which insoluble proteins [amyloid] are deposited outside of cells in various tissues and organs, compromising their normal function); specifically, amyloidosis of the kidney is the deposition of amyloid outside the cells of the blood vessels in the glomerulus (glomerular capillaries), the glomerulus itself, and the tissue spaces (known as “interstitium”)
  - ♦ Nephroblastoma—a congenital kidney tumor
  - ♦ Multifocal renal-cyst adenocarcinoma—a hereditary kidney cancer in dogs
  - ♦ Fanconi's syndrome—a generalized functional abnormality involving the tubules of the kidney, characterized by impaired reabsorption of glucose, phosphate, electrolytes, amino acids, and uric acid
  - ♦ Presence of glucose in the urine due to primary kidney disease (known as “primary renal glucosuria”)—an isolated functional defect in the reabsorption of glucose by the kidney tubules, characterized by the presence of glucose (sugar) in the urine when the pet has normal blood glucose levels
  - ♦ Cystinuria—excessive excretion of cystine (an amino acid) into the urine, caused by an isolated functional defect in the reabsorption of cystine and other dibasic amino acids by the kidney tubules

- ♦ Xanthinuria—excessive excretion of xanthine into the urine, caused by a deficiency in an enzyme, xanthine oxidase, and impaired conversion of hypoxanthine to xanthine and of xanthine to uric acid
- ♦ Hyperuricuria—excessive excretion of uric acid, sodium urate, or ammonium urate into the urine, caused by impaired conversion of uric acid to allantoin by the liver and enhanced excretion of uric acid by the kidney tubules
- ♦ Primary hyperoxaluria—a disorder characterized by intermittent high levels of oxalic acid or oxalates in the urine (known as “hyperoxaluria”), l-glyceric aciduria, oxalate kidney disease (nephropathy), and acute kidney failure
- ♦ Congenital nephrogenic diabetes insipidus—“water diabetes”—a disorder of kidney concentrating ability, caused by diminished kidney responsiveness to antidiuretic hormone, such that the kidney does not reabsorb water and excessive urine is produced

## GENETICS

- Familial (runs in certain families of animals) kidney disorders have been reported in the following dogs and cats:
  - ♦ Renal agenesis in beagles and Doberman pinschers
  - ♦ Renal dysplasia in Alaskan malamutes, boxers, chow chows, golden retrievers, Keeshonden, Lhasa apsos, miniature schnauzers, shih tzu, soft-coated Wheaten terriers, and standard poodles
  - ♦ Glomerulopathy in beagles, Bernese mountain dogs, Brittanys, bull terriers, Doberman pinschers, English cocker spaniels, Newfoundlands, rottweilers, samoyeds, and soft-coated Wheaten terriers
  - ♦ Tubulointerstitial nephropathy in Norwegian elkhounds
  - ♦ Polycystic renal disease in the following breeds of dog: beagle, bull terrier, cairn terrier, West Highland white terrier; and in the following breeds of cat: Persian, exotic shorthair, and Himalayan
  - ♦ Renal telangiectasia in Pembroke Welsh corgis
  - ♦ Renal amyloidosis in the following breeds of cat: Abyssinian, oriental shorthair, and Siamese; and in the following breeds of dog: beagle, English foxhound, and Chinese shar-pei
  - ♦ Renal cystadenocarcinoma in German shepherd dogs
  - ♦ Fanconi's syndrome in basenjis and border terriers
  - ♦ Primary renal glucosuria in Norwegian elkhounds, Scottish terriers, and basenjis
  - ♦ Cystinuria in the following breeds of dog: basset hound, English bulldog, dachshund, French bulldog, Irish terrier, mastiff, Newfoundland, Scottish deerhound, Staffordshire bull terrier, and Australian cattle dog; and in the following breed of cat: domestic
  - ♦ Xanthinuria in the following breed of dog: Cavalier King Charles spaniel; and in the following breed of cat: domestic shorthair
  - ♦ Hyperuricuria in Dalmatians and English bulldogs
  - ♦ Primary hyperoxaluria in the following breed of dog: Tibetan spaniel; and in the following breed of cat: domestic shorthair

## SIGNALMENT/DESCRIPTION OF PET

### Species

- Dogs
- Cats

### Breed Predilections

- Sporadic cases of congenital (existing at birth)/developmental kidney disease can occur without an apparent familial predisposition in any breed of dog or cat; “familial” refers to diseases that run in certain families of animals
- For familial kidney disorders, see “Genetics”

### Mean Age and Range

- Most pets are less than 5 years of age at time of diagnosis

### Predominant Sex

- Familial cystinuria occurs primarily in male dogs
- Samoyed hereditary glomerulopathy is more common in males than females; both sexes are affected in Newfoundlands
- Familial glomerulonephropathy of Bernese mountain dogs is more common in females than males

## SIGNS/OBSERVED CHANGES IN THE PET

- Signs depend on type of kidney disease
- Some pets affected with developmental kidney disease will not have any signs of disease
- Signs related to chronic kidney failure, such as lack of appetite (known as “anorexia”), sluggishness (lethargy), excessive urination (known as “polyuria”) and excessive thirst (known as “polydipsia”), weight loss, vomiting
- Some glomerulopathies are associated with abdominal distension, fluid buildup (known as “edema”), or other signs of the nephrotic syndrome (medical condition in which the pet has protein in its urine, low levels of albumin [a type of protein] and high levels of cholesterol in its blood, and fluid accumulation in the abdomen, chest, and/or under the skin)
- Abdominal distension in some pets with polycystic kidneys or kidney cancer
- Bloody urine (known as “hematuria”) in some pets with renal telangiectasia or kidney cancer
- Apparent abdominal pain in some pets with renal telangiectasia
- Pets with one-sided lack of development of a kidney (renal agenesis), misplaced (ectopic) kidneys, and isolated kidney tubular transport defects are frequently asymptomatic
- Fluid buildup in the abdomen (known as “ascites”) or fluid buildup under the skin (known as “pitting edema”) in some pets with protein-losing glomerulopathies (a group of kidney diseases where excessive amounts of protein are lost through the urine) or amyloidosis (a group of conditions of varied cause in which insoluble proteins [amyloid] are deposited outside of the cells in various tissues and organs, compromising their normal function)
- Enlarged kidney (known as “renomegaly”) or abdominal mass lesions may be identified in some pets with polycystic kidneys, kidney cancer, or fused ectopic kidneys
- Renal pain may be seen in some pets with renal telangiectasia

## CAUSES

### Non-hereditary

- Infectious agents—feline panleukopenia virus and canine herpesvirus infection have been associated with renal dysplasia
- Drugs—steroids, diphenylamine, and biphenyls have been associated with polycystic kidneys; chlorambucil and sodium arsenate have been associated with renal agenesis
- Dietary factors—too low levels of vitamin A (known as “hypovitaminosis A”) or too high levels of vitamin A (known as “hypervitaminosis A”) have been associated with renal ectopia

## RISK FACTORS

- See factors listed under “Causes”

## Treatment

### HEALTH CARE

- The nature of congenital and developmental kidney disorders often prevents specific treatment
- Supportive or symptomatic treatment may improve quality of life and minimize progression in pets with kidney dysfunction
- Treatment options are based on clinical signs and laboratory diagnostic testing
- Specific treatment is determined after diagnosing the type of kidney disease or clinical syndrome

### DIET

- Pets with chronic kidney failure—restrict phosphorus and moderately restrict protein
- Pets with high blood pressure (known as “hypertension”)—restrict sodium

## Medications

Medications presented in this section are intended to provide general information about possible treatment. The treatment for a particular condition may evolve as medical advances are made; therefore, the medications should not be considered as all inclusive

- Specific treatment is determined by diagnosing the type of kidney diseases or clinical syndrome

## Follow-Up Care

### PATIENT MONITORING

- Specific monitoring is determined by diagnosing the type of kidney diseases or clinical syndrome
- Bloodwork (such as complete blood count, serum biochemistry profile) and urine testing (such as urinalysis, microalbuminuria testing, urine protein:creatinine [UP/C] ratio)

## **PREVENTIONS AND AVOIDANCE**

- Congenital and developmental kidney disorders are irreversible, so control lies in preventing breeding of affected pets
- Always consider early identification and correction of predisposing factors (genetic and non-genetic) that may affect future offspring

## **POSSIBLE COMPLICATIONS**

- Acute or chronic kidney failure
- Nephrotic syndrome (a medical condition in which the pet has protein in its urine, low levels of albumin [a type of protein] and high levels of cholesterol in its blood, and fluid accumulation in the abdomen, chest, and/or under the skin)
- Presence of stones in the urinary tract (known as “urolithiasis”)
- Dilation of the cup-shaped cavity or pelvis of the kidney due to blockage of the flow of urine (known as “hydronephrosis”)
- Urinary tract infection

## **EXPECTED COURSE AND PROGNOSIS**

- Highly variable; depends on the specific disorder, the extent of primary lesions, and the severity of kidney dysfunction
- Most congenital (existing at birth) and developmental disorders are irreversible and may result in advanced chronic kidney failure, but some pets with mild-to-moderate kidney dysfunction may remain stable for long periods
- Pets with some disorders may remain without signs of disease (known as being “asymptomatic”), unless the disorder is complicated by development of urinary stones (urolithiasis), urinary tract infection, or other disease processes that promote progressive kidney dysfunction

## **Key Points**

- Congenital (existing at birth) and developmental kidney diseases are a group of diseases in which the kidney may be abnormal in appearance or may be abnormal in its ability to function normally or both
- These diseases result from inherited or genetic problems or disease processes that affect the development and growth of the kidney before or shortly after birth
- Congenital and developmental kidney disorders are irreversible, so control lies in preventing breeding of affected pets
- Supportive or symptomatic treatment may improve quality of life and minimize progression in pets with kidney dysfunction

# Notes

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