

# Cystinuria

## BACKGROUND

Cystinuria is an inherited, metabolic disorder. The affected individual has an impaired capacity for reabsorption of the amino acid, cystine, in the kidney tubules. Because cystine is not reabsorbed by the kidneys, it becomes a part of the kidneys' product: urine. Therefore, cystinuric individuals have an excess of cystine in their urine. Most of the time, this excess cystine is simply excreted in the urine. Because cystine is one of the sulfur-containing amino acids, the urine may have a characteristic "rotten egg" odor.

As long as cystine stays in solution in the urine, it will be excreted without incident. It is when excessive cystine in the urine is not excreted that it becomes problematic. Sometimes, cystine will precipitate, or solidify, forming stones. You may also see stones referred to as crystals, uroliths, or calculi; these are interchangeable terms. Cystine stones can cause problems for the person or animal affected, by blocking the urethra, bladder, or kidney. This results in prevention or slowing of urination. There may be straining to pass the urine, and there may be blood in the urine. Some individuals will pass the actual stones, if they are small enough to be passed instead of causing blockage. In severe cases, the blockages can lead to rupture of the bladder or kidney, which may even be fatal.

## TREATMENT

There are three methods of control for cystinuria.

Medical management is a possibility, through the administration of drugs that will help to dissolve the stones. The drugs will not prevent the stones from forming; they will only assist in "shrinking" already formed stones. Drugs also come with side effects, and may not be a wise course of treatment for long term use.

Surgery may be indicated, in certain individuals. Usually it has been found that the stones removed during surgery will eventually reform later on, necessitating further surgery or some other method of treatment.

Dietary modification, with the accompanying alkalization of the urine, is the approach I recommend. The intent of this approach is to reduce the likelihood of the stones forming in the first place. This way, there is no need to surgically remove or medically shrink anything! It may seem that if we simply avoided feeding anything with cystine in it to the animal, that would solve the problem. After all, if there were no cystine, we would not have to worry about it being reabsorbed. Unfortunately, that is not the case. Cystine is not the "enemy," in fact, it is an essential amino acid, meaning it must be a part of the diet of a healthy individual. It is so necessary, that the body will actually manufacture

cystine out of other amino acids (methionine) if the diet is deficient in cystine. So, we will always have to be concerned with how best to rid the body of the cystine that cannot be reabsorbed. Of course it also makes sense that feeding the lowest cystine-containing sources of foods is a smart thing to do; we just can't expect to eliminate cystine from the diet completely.

## GOAL OF DIETARY MODIFICATION

There are many types of urine stones that can develop in people and animals. Cystine stones behave differently from other types of urinary stones (for example, struvite) in that they tend to form when their environment (the urine) is acidic (low pH). Therefore, the goal in modifying the diet of an affected individual is to alkalinize, or raise, the pH of the urine. This creates an environment where the excess cystine will be less likely to form into stones, and more likely to be excreted as part of the urine solution.

How can we control the pH of the urine? Well, the pH is largely influenced by the diet consumed. Generally speaking, high-protein diets, based on large quantities of meats, lead to acidic urine. Low-protein diets, based more on plant materials, lead to alkaline urine. The ideal diet is designed to balance the proportions of meat and vegetables, such that the dogs' requirement for protein is met, but not vastly exceeded.

Another way to trick the urine into becoming more alkaline is to use what are called buffering agents. These are substances that will react in the digestive system and internal organs, to de-acidify, or "buffer" the urine components. This results in maintaining the pH at a higher level, thereby preventing stone formation. The most common buffer is plain old baking soda (sodium bicarbonate). Some special forms of vitamin C also serve to help control the urine pH.

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## CONCLUSION

It is important to understand, as best we can, what is going on inside the bodies of our pets. This enables us to provide the best care we know how to give to them. Cystinuria can be controlled and managed; it need not mean illness or early death for your dog. It is not recommended to breed affected dogs, since this is a genetic condition. There is current research being conducted on cystinuric dogs, in an attempt to map the genes responsible and perhaps find a way to prevent its inheritability one day in the future.

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