

# Fanconi Disease Management Protocol for Veterinarians

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**DEFINITION:** FANCONI DISEASE is a mammalian renal tubular reabsorption failure, usually genetic in origin, resulting in polyuria/polydipsia (PU/PD) with glucosuria; generalized aminoaciduria; proteinuria; loss of multiple vitamins, minerals, electrolytes and bicarbonate. These losses result in metabolic acidosis, protein-loss muscle wasting, weight loss, and myalgia. Left uncorrected, it causes increasing acidosis, while stressing the respiratory compensation system to its maximum effort, and leads to the progressive and degenerative renal, and then multisystem failure, resulting in death. With early intervention and lifelong management, prognosis is excellent for long term, healthy survival. Very compliant dogs have lived to normal lifespans. Correction of the acid/base component appears to slow dramatically the degenerative progression. This disorder is prevalent in the Basenji breed of dog, although it appears in many breeds of dog and in other mammals, including humans. There is also a similar syndrome now identified in birds, but I do not know if this Protocol is applicable to avian physiology. This Protocol has been used in a number of other breeds of dog, with slight increases in bicarbonate doses for larger dog breeds being the only change. It has also been applied successfully to a Fanconi afflicted horse and several cats.

**DIAGNOSIS:** All Basenjies should be tested by their owners **monthly** for **urine glucose**, using Ames Combistix, or any similar test strips / tape, available from the diabetic testing section of pharmacies. Any positive reading should be reported to their veterinarian. Disease often presents as polyuria/polydipsia or UTI. Positive diagnosis is made by glucosuria with normal or low blood glucose. In Basenjies, Fanconi is considered genetic. In other breeds, INDUCED Fanconi should be considered and vectors ruled out, with toxins removed from environment to help prevent further renal damage. Induced Fanconi in ANY breed, including Basenjies, can be self-limiting, but is treated the same as genetic Fanconi, with correction of deficiencies. Most common misdiagnosis is diabetes, or Cushing syndrome **ONSET:** Age of first symptoms (PU/PD) is usually 5 to 7 years; however, it has been diagnosed from 3 to 11 years. Early diagnosis is essential. The earlier treatment is begun, the less renal damage exists, and thus fewer replacements are needed.

**LABORATORY TESTS TO RUN: Initial Diagnosis and Quantification of Fanconi-induced losses:** Once a working diagnosis of Fanconi disease has been made on the basis of a POSITIVE urine glucose in the absence of elevated blood glucose (and without predisposing events, such as poisoning or renal infection), then the following tests should be run:

1. **Venous Blood Gas Panel** (looking for acidosis, low PaCO<sub>2</sub>, low bicarbonate level and low Base excess). These labs can usually be run by a human hospital, or new, inexpensive monitors like the I-Stat, and are **ESSENTIAL** (total CO<sub>2</sub> on a chemistry panel **does not** suffice). Further specifics of blood gas analysis follow later in this protocol. Be aware that when the dog is in respiratory compensation mode, the CO<sub>2</sub> level can drop very low, without visually evident hyperventilation or tachypnea. This is why the CO<sub>2</sub> on a blood gas panel is often such a vital tool to assess the severity of the disorder. In very "late" cases, the respiratory compensation mechanism will have failed and the CO<sub>2</sub> will be trending up to normal, while the pH is dropping precipitously, thus sending the dog into a potentially fatal acidosis crisis. Aggressive management in these "late" diagnosed cases may still be life saving. IV bicarbonate and even fluid dialysis have proven life saving. While this Protocol only lists CO<sub>2</sub> and pH as axis to follow to dose bicarbonate, HCO<sub>3</sub> and Base Excess MUST be considered. Our goal is always to maintain a blood level of 20 or greater on the venous blood gas HCO<sub>3</sub>. This is measured 8 hours after last bicarbonate dose in a managed dog.
2. **Serum multi-chemistry panel**, especially **sodium, potassium, calcium, and phosphorous** (looking for deficiencies).
3. **Thyroid Screening.** A fair percentage of Fanconi afflicted dogs have been shown to be hypothyroid. I would suggest waiting until the first Fanconi follow up (Protocol in place) before performing a T4, since "out of control" Fanconi can cause false T4 readings. Even if the thyroid screen shows hypothyroid, and replacement therapy is instituted, you should recheck a level in six months, after a Fanconi treatment regimen is begun.
4. **Urine Screening.** Afflicted dogs will ALWAYS have dilute and alkalotic urine, from the high volume of ingested water and the bicarbonate loss. Likewise they will always remain glucosuric. What we want to check for on EACH follow up visit, is the presence of any possible UTI, since the sugar and pH situation in the bladder and kidney is a perfect set up for infection. IF ANY SYMPTOMS SUGGEST UTI, EVEN WITHOUT POSITIVE FINDINGS VIA URINALYSIS, it may be a loculated micro-infection, and many have benefited from a single run of antibiotics. Recurrent UTI in a Fanconi dog requires a urine culture, since any number of infections may occur in an altered state of resistance, caused by systemic, chronic illness.

**FOLLOW-UP LABORATORY SUGGESTIONS:** Years of experience have shown that a good level of success can be achieved by following these basic guidelines for Fanconi-related lab studies:

1. INITIAL BLOOD GAS and CHEMISTRIES as above noted.
2. Follow up, usually eight to ten weeks after beginning full management regimen. This allows time for compensation mechanisms to slow down or stop, thus giving accurate long-term lab values.
3. Repeat follow up, with all labs at 6 months after first follow up.
4. Annual checkups and labs if all is going well. (Although some vets prefer six month follow-ups, at most).
5. In repeat labs, try to make sure that the time interval between bicarbonate and vitamin dosing and blood being drawn is consistent with each recheck. If bloodwork is to be done in the AM, then no morning pills should be given; likewise, afternoon blood draws should be done late enough to avoid the peak absorption period. We want to try and get a set of labwork that approximates where the dog is existing most of the time, between pill doses. Eight hours after last bicarbonate and vitamin dose gives a good idea of "trough" level blood chemistry.

Most dogs, once controlled, are stable and healthy enough to follow this schedule for lab and doctor visits. **MORE frequent follow ups or labs are NEVER an objection**, if justified by the veterinarian's clinical intuition or if the dog is, in **any way**, showing less than extreme good health and optimal control of their disease. Also, any acute illness, pending surgery or other unusual stress is ample justification for repeating labwork.

**THERAPEUTIC GOALS:** Restoration of normal blood chemistry by matching losses of bicarbonate, protein and vitamins/minerals. Maintain body habitus and restore good muscle tone and activity level.

1. Bicarbonate is dosed, using blood gas analysis, to match the Fanconi induced losses and return blood to normal acid/base balance without depending on body's short-term compensatory mechanisms. Success will be seen as an increasing PCO<sub>2</sub> (as the respiratory compensation mechanism, which demand metabolic work, turns off), and increasing HCO<sub>3</sub> levels.
2. Create a positive protein balance allowing return to tissue building and to support muscle mass and strength. This is accomplished with a HIGH protein diet, since ordinarily this is NOT "renal failure." In cases compounded with renal failure, such as other acute diseases or chronic changes of old age, see modification of the protocol mentioned later, under "Renal Failure "Hybrid" Protocol."
3. Normalize Electrolytes. Support the body's vitamin/mineral needs, including trace minerals. Note: Correction of bicarbonate loss and correction of blood chemistry seems to slow progression of this disease to the point that it is inconsequential in a dog's life span. Serial GFR and creatinine clearances in some research test dogs have confirmed this, although these tests are too expensive and invasive to recommend routinely. Acute one time dialysis is rarely needed, but has been used successfully as a lifesaving measure when late diagnosis has resulted in acute renal failure. Kidney function has returned to post-dialysis in some of these cases.

**MANAGEMENT DOSING GUIDELINES:** Note that these are recommendations only, based on almost fifteen years and many hundreds of dog's experience. NO general guideline can take into account all the variables that any individual condition may present, thus we ask that ALL veterinarians use their individual clinical training and judgment in prescribing and designing any treatment regimen for an afflicted dog.

**This protocol is designed only as a tool to assist the veterinarian and should NEVER be used without the care and expert advice of a veterinarian in treating any dog. THIS IS A MUST!!!**

**INITIAL MINIMUM TREATMENT FOR POSITIVELY DIAGNOSED DOGS-INDIVIDUAL DOG'S LOSSES WILL BE CORRECTED BY TITRATING UP FROM HERE.** Please note that all dosages are based on the average 22-27 lb. (10-12.5 kg) Basenji. Please calculate doses up or down for treating other size dogs or animals afflicted with Fanconi.

- 1. Fresh water freely available.** Do not add any medication or supplements to regular water or food items as this may cause a dog to stop eating their regular meals. Supplements and medicines should be given as a separate "treat" offered twice a day.
- 2. Any good quality dry food** may be fed, with the addition of at least one can per week of HIGH PROTIEN "wet meat" mammal meat based dog food (Beef, Lamb...etc.) to replace long-chain amino acids and phosphorus. Poultry based foods have proven far less effective in management of this disorder than mammal based meats. The only exception to this high protien diet, in this protien-losing uropathy, is documented renal failure (increasing BUN/CREATININE). In this type situation, see the "RENAL FAILURE HYBRID PROTOCOL" section later in this instruction set.
- 3. PET-TAB PLUS** type vitamin/MINERAL supplement tablet. ONE tablet, divided ½ tab BID for **asymptomatic** dogs diagnosed ONLY by positive glucosuria. TWO tabs daily, given ONE TAB BID for **symptomatic** dogs. TITRATE HIGHER for dogs with MINOR hypokalemia or hypocalcemia. Make sure any supplement has MINERALS, not just vitamins.
- 4. PET-CAL or CALCIUM PHOSPHORUS (By PET-AG)**-type vitamin D-phosphorus replacement tablet. Given ½ tab BID in asymptomatic dogs. ONE tab BID in symptomatic dogs. Another option is **Dicalcium Phosphate powder**, a "Horse Health" product that also lists dog doses on the package. One-teaspoon equals one Pet Cal tablet. Some dogs like the taste better, and it is less expensive than tablets. Phosphorus loss is a hallmark of Fanconi disease and since it is sequestered from muscle mass into the blood, standard chemistry panels may not reveal the full extent of the loss. IF Fanconi afflicted dogs show a loss of muscle mass and any sign of MYALGIA after they appear corrected on BLOOD GAS and CHEMISTRIES, increasing the PET-CAL and AMINO FUEL may help. Pet Cal type vitamin should be stopped in renal insufficiency cases, to decrease phosphorus.
- 5. CENTRUM VITAMIN**-type COMPLETE vitamin/mineral tablet (high potency), at a dose of ONE tab/week in dogs with PU/PD (no dose needed in fully asymptomatic dogs). This covers the loss of many TRACE elements caused by the high water washout of the PU/PD). In cases of "unusual symptoms" in a corrected dog, such as seizures, acute onset blindness or other problems without clear cause, it does not hurt to empirically TITRATE UP the CENTRUM to as high as one tab EVERY OTHER DAY, since we have seen multiple "strange" symptoms resolve this way. Trace elemental losses and deficiencies in these dogs may have well caused these symptoms.
- 6. AMINO FUEL ("Stack" by Twinlabs)**-type COMPLETE amino acid preparation. (Tablets available in body building section of General Nutrition Center or other similar store). Dose at one tab/week if asymptomatic. Titrate up as high as one tab, every-other-day, in cases of extreme muscle wasting, poor coat or unresolved skin problems. In cases where a HYBRID renal failure/Fanconi regimen is being used, the Amino Fuel dose goes as high as ½ tab BID (when given with LOW protien foods to correct increased BUN and CREATININE). Amino acid body building pills come in many "dose ranges", the LOWEST DOSE "Complete" formula you can find is perfect.
- 7. SODIUM BICARBONATE ANTACID TABLETS- (Brands include URL, Rugby, Lily, Watson and Concord.)** This is THE MOST IMPORTANT component of this protocol. Without correction of the Fanconi bicarbonate loss and correction of serum acid/base imbalance, this disease remains fatal. Sodium Bicarbonate 10-grain antacid tablets (similar in size and appearance to an aspirin tablet) are available OTC from any major pharmacy. It is very inexpensive and thus most economical to purchase in a 1000 count bottle form. (Current 2003 pricing varies from \$12 (Wal-Mart Pharmacy) to \$30 per 1000 tablets at national drugstore chains). I do not recommend using powdered bicarbonate (baking soda), since the volume of dosing would be very difficult and the level of medical purity (compared to the tablets) is questionable. Bicarbonate will be dosed based upon a VENOUS BLOOD GAS PANEL. Emergency dosing can be done at a rate of THREE 10 grain tablets BID, but this is VERY undesirable compared to a measured correction. **BICARBONATE TABLETS MUST BE GIVEN INTACT.** Pills can be hidden in a small amount of food, such as VELVEETA cheese ball or baby food ball or hidden in a bit of meat...etc Some dog respond well to GENTLE "Pilling" and there are devices to help. Others do much better with "Treat-time"...with LOTS of excitement, seems to make the twice a day pill giving time a lot easier on owner and dog. In cases where the dogs are resistant to taking the bicarbonate, it can be crushed up (the least amount possible) and then placed inside hollow gelatin capsules (available at pharmacies and health food shops). These capsules can then be hidden (without as easy detection by the dog) in a little treat for administration. Also, it has been reported that some dogs get "gassy" or flatulent with the bicarbonate, but this passes in a few weeks. Meanwhile, some dogs tolerate the bicarbonate best at mealtime (less apparent GI distress). Other dogs do much better when the bicarbonate is spaced away from their meal time by an hour or two, so that the normal stomach acids are not neutralized while in the presence of food, which can result in vomiting and poor food tolerance. This finding will vary from dog to dog, so trial and error is the best way to optimize the pill and food relationship. NOTE: For European users. A **10 Grains** tablet of Sodium Bicarbonate equals **650 mg**. PLEASE NOTE, CITRATES, like UroCit K and others, HAVE NOT WORKED at buffering the blood in acidotic Fanconi dogs. It is FINE for replacement of potassium, but has proven useless in multiple trials, at restoring normal blood pH in Fanconi dogs. It may work in other disease processes, but for Fanconi. We have found NO buffer that works as well (or at all) compared to replacing the lost Bicarbonate WITH bicarbonate. This is why this Protocol, as multiple human Fanconi Protocols do, stresses the importance of BICARBONATE REPLACEMENT as a key element.
- 8. POTASSIUM SUPPLEMENTATION** such as TUMIL-K (2 MEQ/Tab) or UroCit-K (5 MEQ/Tab) or over the counter "Nature Made" brand 550-mg Potassium Gluconate tablet is used in about 5% of Fanconi dogs for persistent hypokalemia, even once otherwise "corrected." These tablets are dosed by blood chemistry and dogs taking them should be followed a bit more frequently. These tablets must also be given intact, especially UROCIT-K, which is a "timed release matrix" delivery vehicle. Crushing some potassium tablets can result in catastrophic overdose for the dog.

**POTASSIUM DOSE RECOMMENDATION SCALE:** Note that TUMIL-K is 2 MEQ per tablet. Nature Made brand is about 2.3 MEQ per Tablet **Potassium**

Measure in Blood	Recommended Starting Dose of Potassium
1.50 to 2.00 MEQ/L	15 MEQ (1620 mg) P.O. B.I.D.
2.10 to 2.75 MEQ/L	10 MEQ (1080 mg) P.O. B.I.D.
2.76 to 3.75 MEQ/L	5 MEQ (540 mg) P.O. B.I.D.

As with any administration of potassium, repeat lab work should be followed closely until stable (one lab per week for four weeks, and as symptoms dictate). Thereafter, routine follow-up should suffice, but bloodwork should be done at six month, rather than one year intervals. No cases of hyperkalemia have been encountered to date, but titrate up carefully

**ACUTE EMERGENCY MANAGEMENT:** fluid has been used successfully as a lifesaving measure when late diagnosis has resulted in acute renal failure. Kidney function has returned to pre-dialysis levels in some cases. Calculating volumes for Fluid dialysis must account for these dogs' already high fluid intake and losses. 3X base fluid levels have been used successfully. Adding I.V. bicarbonate to slowly correct pH. and I.V. nutritional support "if available" may help with life support and recovery. Oral alimentation once eating (Sustical, Nutrical, or Ensure) should be used till the dog is weaned off IV to regular diet.

**RENAL FAILURE "HYBRID" PROTOCOL:** As more dogs have survived long term on this protocol, we have seen multiple cases of renal failure onset from various causes, most often just the "normal" slow onset renal failure of aging. In these cases we have HYBRIDIZED the Fanconi Protocol to allow for the maintenance of acid/base chemistry, while correcting the problems of the renal deficiency. We start with life saving measures, including fluid, or peritoneal dialysis as needed. We then institute a diet as follows:

1. LOW protein dry or "canned" food. Fresh water remains freely available.
2. Add in up to ½ tablet of the AMINO FUEL or other amino acid preparation daily, to cover our protein loss.
3. Increase CENTRUM type multivitamin to QOD, or use a human COMPLETE formula for MEN (some have no phosphorus).
4. Drop the PET-CAL type tablet from the regimen, since in renal failure serum PHOSPHORUS tends to go UP.
5. Labwork and physical exams should be performed MORE frequently in renal insufficient dogs.



**OTHER LAB VALUES TO NOTE:** In many medically managed Fanconi dogs, we have seen some unusual chemistry elevations. Some of these values would imply liver disease, but thus far, it not been the case, and these dogs have shown no symptomology or problem associated with these values. Some researchers have speculated that these abnormalities are "normal" in a Fanconi dog due to altered blood density from protein loss. This causes shifts in certain enzymes and blood components, resulting in these elevations. Of course, any changes must be followed, since the PRESENCE of Fanconi does NOT rule out the possibility of developing liver disease, or any other unrelated problem.

Lab Test Results	Normal Value	Elevated Values we have seen in Fanconi patients
Alkaline Phosphatase	20-200 IU/L	200 - 850 IU/L
SGOT	25-105 IU/L	105 - 310 IU/L
SGPT	10-75 IU/L	75 - 500 IU/L
Cholesterol	137-275 IU/L	275 - 450 IU/L
Triglycerides	20 - 80 mg/100 ml	80 - 900 mg / 100 ml

Of course, the dog's last meal and other factors can also affect these values. They are worth noting, but usually stabilize at some level and may not need treatment. If you choose to treat, use caution. For instance, one would not want to lower cholesterol at the expense of depriving protein, especially in a protein-losing disorder.

**HELPFUL HINTS IN SEVERAL AREAS: MEDICAL MANAGEMENT:** Periodic follow-up for UTI is essential since there is always a glucosuria present. Antibiotics appear to be tolerated normally. While symptoms such as PU/PD should be watched, these dogs have tolerated short-term steroids, via I.M., I.V., and P.O. routes for neurologic and orthopedic problems. Chronic but periodic low dose steroid use for treating flea induced skin problems has also appeared to be benign. Chemotherapeutic agents have been used on some Fanconi dogs for treatment of different tumors. Their reactions were not observed to be different than the non-Fanconi canine population, although the less nephrotoxic agents have been the choice of the oncologist involved. Some dogs have been mildly incontinent even with correction of all measurable values. In these cases, **Phenylpropanolamine HCL (PPA)** Orally chewable "Proin" by "Phamacal" twice a day, last dose one-hour before bed, has allowed owners and pets to sleep through the night and prevent bed-wetting. The PPA strengthens the bladder sphincter valve, and can assist in Fanconi afflicted, where urine volume, glucose and pH all contribute to potential "urgency" or "leaky" incontinence. **Please use this medication only with a fully corrected dog, since it only MASKS symptoms. I would prefer to correct any underlying problem first.** PPA dosing is usually 1-2 mg/kg, q 12 hours. Sudden change in urination habits and any incontinence in the Fanconi dog should first be considered as a urinary tract infection, until proven otherwise.

**SURGICAL MANAGEMENT:** General anesthetics are well tolerated. Pre-anesthetic K+ level is advisable. Emergence from general anesthesia, as well as the reversal of sedative hypnotics, appears slightly prolonged. I strongly suggest that emergence from general anesthesia be accompanied by supplemental oxygen, as weakness or "hypoventilation induced hypoxia" in these already compromised dogs presents unneeded stress. In prolonged NPO states or during long surgery, several MEQ of HCO<sub>3</sub> (I.V. form) may be added to a liter of IV fluid can help prevent intraoperative systemic acidosis. (25 MEQ per 1000 cc of IV fluid)

**GENERAL INFORMATION** A controlled Fanconi dog may be treated as any other dog. They are prone to any other disease process and are able to be medicated as any other dog. Exercise **should not** be limited, (There are runners, mountain climbers and lure coursing champions who are well-managed Fanconi patients) although **fresh water should always be available**, and afflicted dogs should be offered water **much more often** than "usual" during such stress activities. Ideas for pill hiding treats include, Velveeta Cheese Slices, which have a malleable, Play-Doh type consistency and can be "formed" into balls with pills inside. Large elbow macaroni cooked in beef or chicken soup or stock (prepare in advance and refrigerate). A few noodles are given as a tease, then give the pill filled ones. Hiding the pills in tiny balls of beef baby food or cat food (more aromatic than dog food), hot dog slices, or bratwurst has also worked. Small quantities ice cream, cream cheese, peanut butter to bologna have been used by various owners. Creativity in this is vital, but remember, once you find a pill hiding technique that works, stick with it as long as the dog does. These are creatures of habit. Owners of multiple dogs can use competition / jealousy between dogs to make pill giving easier, making sure the non-afflicted pets are given a NON-medicated treat, while avFanconi dog is given its pill-hiding treat. Competition makes the pill-containing treat seems to get swallowed faster and without much "investigation".

**ACTIVE DOG MANAGEMENT:** With some Fanconi dogs actively lure coursing, hiking or doing other vigorous activity, care must be taken to provide adequate water to meet dogs needs. Also, avoid overheating a Fanconi dog. Finally, an extra bicarbonate tablet before and after hard exercise can help compensate for lactic acidosis from muscle use.

**Granulomatous Meningoencephalitis (GME).** If a Fanconi dog exhibits blindness, lameness or any other "odd" neurological symptoms, GME should be considered. This is a NON contagious, **multi-site brain and spinal cord tumor**. There has been a noticeable incidence of GME in Fanconi dogs. The link is not known, and NOT all Fanconi dogs develop GME. It is best diagnosed with a CT Scan with Hypaque type contrast. MRI can also be used. No known treatment has been successful for GME. Dexamethasone or Prednisone can offer some symptom relief and slight increase in life-span, but GME is an aggressive, Fast growing cancer. This is a dose schedule that has appeared on several veterinary medical websites:  
Dexamethasone 0.25mg TID x 7 days, BID x 21 days, SID x 42 days, EOD x 42 days.

**FURTHER INFORMATION ON FANCONI:** Many wonderful sites are available on the INTERNET with suggestions on "pill hiding treats" and other topics pertaining to owning and feeding a Fanconi afflicted dog. Likewise, many local or regional Basenji breed club magazines have interesting owner articles on this subject. **My motto is that information is always your best ally in fighting any disease.** A great place to start is with nationally circulated breed magazines or with some of the excellent regional and breed club newsletters. "Networking" with other owners via clubs and shows is a great asset to "staying in touch" with pertinent health news and breakthroughs. There are some online support groups which can really help as well.

#### **SUGGESTED SITES and PERIODICALS:**

I would STRONGLY suggest reading one, or even ALL THREE regularly. They are entertaining and VERY importantly educating on the latest Basenji health issues. Even NON Basenji Fanconi animal owners can gain a LOT by reading and following the progress in these locations.

BASENJI CLUB OF AMERICA (BCOA) (Club website and magazine)- <http://www.basenji.org>  
BASENJI COMPANIONS (website and newsletter-magazine)- <http://www.basenjicompanions.org>  
THE BASENJI MAGAZINE (magazine and website) -<http://www.thebasenji.com>

#### **CONTACT NUMBER, IF NEEDED.**

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**(912) 598-5067 please limit calls to the hours of 7:30 pm to 9:00 pm EST** or e-mail is always welcome at: [Outdoc@aol.com](mailto:Outdoc@aol.com)  
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