Six months ago this 23-year-old electrical engineering student began having difficulties completing his drafting assignments and started fighting with his roommate. Three months later, when criticized for doing sloppy work and damaging some delicate experimental equipment, he responded with a string of profanity. He was seen to be uncoordinated and tremulous, his speech was slurred, and he was making grotesque faces. He was dismissed from class for being drunk and disorderly. His course since then has been progressively downhill. His past medical history is negative but for anemia, discovered during a camp physical examination 10 years ago. When brought to the emergency room by his roommate, he was drooling saliva, had uncontrollable hands, head, and body tremors and muscular rigidity. He seemed demented. The eye findings are pathognomonic.

His blood urea nitrogen was 24, creatinine 1.7 mg/dl, sodium 140 mEq/l, chloride 104 mEq/l, potassium 4.0 mEq/l, bicarbonate 26 mmol/l, phosphorus 1.6 mg/dl, uric acid 3.7 mg/dl, and glucose 95 mg/dl. His urine-specific gravity was 1.014, pH was 5.3, proteinuria was 2+, glucosuria was 1+, 24-hour urine protein was 1.4 g. Creatinine clearance was 38 cm³/min, 24-hour uric acid 556 mg, phosphorus 2.7 g, calcium 424 mg, and glucose 2.2 g. His serum ALT was 20 U/l, AST 22 U/l, and GGT 51 U/l. Bone x-rays showed subarticular cysts, fragmentation of bone around his wrists, and vertebral osteo-chondritis. Questions:

What is the diagnosis?
What are the most definitive tests to confirm the clinical diagnosis?
What is the treatment for this condition?
What is the prognosis without treatment?
What is wrong with his kidneys?
What is wrong with his bones?

A 39-year-old white woman who had been told she had analgesic nephropathy was given tetracycline in a dosage of 500 mg per os t.i.d. for 5 days by a physician at a walk-in clinic who did not have access to her medical records for the treatment of symptoms compatible with an upper respiratory infection. Her serum creatinine level for the previous 6 months had varied from 4.5 to 5.5 mg/dl, and her blood urea nitrogen level between 62 and 77 mg/dl. Six days after starting tetracycline, she complained of epigastric pain, nausea, vomiting and weight loss. When admitted to the hospital, she was in acute distress due to the abdominal pain. Her blood pressure was 106/66 mm Hg (supine) with a pulse of 96, and 84/50 mm Hg (standing) with a pulse of 128; her temperature was 38 °C. She complained of epigastric tenderness without rebound or ileus. The rest of her physical examination was unremarkable. Serum creatinine was 16.5 mg/dl and blood urea nitrogen 422 mg/dl. The initial serum amylase was 1,536 U/l (normal level: 60–
Electrolytes were as follows: sodium 148 mEq/l, chloride 90 mEq/l, potassium 3.6 mEq/l and CO2 20 mmol/l. Liver function was normal. WBC 11,000/mm3, 88% segs., 6% bands, 6% lymphocytes. Hematocrit was 41 % and hemoglobin 9.6 g/dl. Urinalysis revealed no evidence of infection or hematuria, urine sodium was 72 mEq/l, chloride 30 mEq/l, and potassium 25 mEq/l.

The patient was treated for acute pancreatitis and hemodialysis was initiated. The pancreatitis resolved after 5 days of treatment; however, her renal function did not improve. After 3 weeks, her creatinine was still in the range of 16 mg/dl and blood urea nitrogen was 84 mg/dl. Radiographic studies included an ultrasound of the abdomen, and kidneys showed small kidneys unchanged from a previous examination without evidence of obstruction. An upper gastrointestinal series was normal. She was discharged from the hospital and continued on maintenance hemodialysis as an outpatient. After 3 months, her renal function still had not improved (creatinine 15–16 mg/dl predialysis). Questions: What was the cause of the pancreatitis?