# A Tale of Two Drugs
# Adverse Anesthetic Reaction in Cystinuria

Nadezhda Bulekova [1], Haleh Saadat MD FAAP [1,2,3]

Frank H. Netter MD School of Medicine at Quinnipiac University, CT,[1] Hartford Healthcare [2] Department of Anesthesiology, St. Vincent's Medical Center [3]

## Introduction

**Definition and Epidemiology:**
- An inherited disorder causing recurrent cystine kidney stones.
- Affects 1 in 7,000 to 10,000 people globally.

**Genetic Background:**
- Mutations in SLC3A1 and SLC7A9 genes.
- Autosomal recessive inheritance.

**Pathophysiology:**
- Poor reabsorption of cystine in kidneys.
- Cystine crystallizes to form stones.

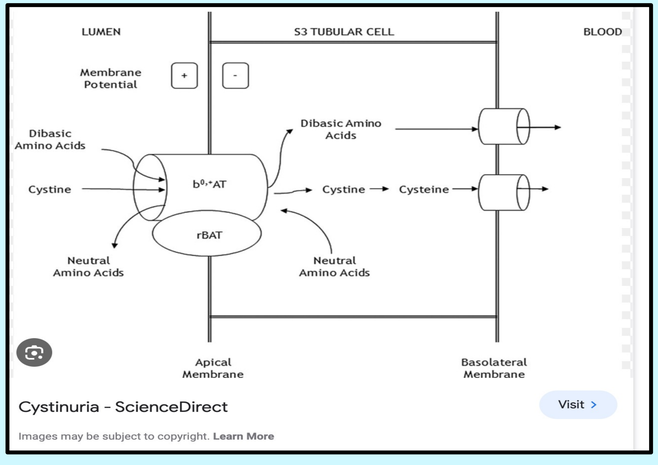
**Clinical Manifestations:**
- Varies from childhood to adult kidney stones.
- Pain, blood in urine, infections.

**Diagnosis:**
- Genetic tests or urine cystine crystals.
- 24-hour urine test for cystine.

**Current Management Strategies:**
- High fluid intake, diet changes,
- Medication.


## Case Report

- A 38-year-old female with cystinuria for cystoscopy
- Multiple previous cystoscopies were uneventful
- Event: Post-succinylcholine administration led to prolonged intubation-Pseudocholinesterase deficiency
- Next Procedure: The anesthesia changed to Rocuronium
- The patient still experienced a similar adverse response (prolonged intubation)
- Further review revealed the patient's use of Thiola for cystinuria
- **Thiola may induce anti-acetylcholine receptor antibodies, potentially mimicking myasthenia gravis symptoms**




## Discussion

- Thiola (tiopronin) is used in cystinuria
- Reduces urinary cystine levels by binding with cystine
- It increases the solubility of Cysteine
- Tiopronin is a thiol compound with antioxidant properties.
- It may bind to heavy metals.
- It can potentially induce anti-acetylcholine receptor antibodies.
- Side effects may mimic myasthenia gravis (MG), affecting nerve-muscle junctions.
- Requires careful consideration in anesthetic management and surgery planning.

## References

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