

QUIZ


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1. A 42-year-old male with severe, recurrent cystine nephrolithiasis has been on maximum tolerated doses of tiopronine, high fluid intake, and consistent urinary alkalization for the past year. Despite this, he continues to form multiple small cystine stones. Which of the following strategies might be considered as the NEXT step in his medical management?

- A. Switching to penicillamine.
- B. Adding a thiazide diuretic to the regimen.
- C. Increasing the dose of potassium citrate to achieve a higher urine pH.
- D. Initiating extracorporeal shock wave lithotripsy (ESWL) for stone burden reduction.

ANSWER A

- A low-protein (< 20 g/day), low-salt (< 2 g/day) diet with high hydration (> 3 liters/day) is strongly advised in children with cystinuria.
- Cystine stone formation can be reduced by increasing the urinary pH level. The solubility of cystine does not increase significantly until a urine pH level above 7-7.5 is reached.
- Cystine-binding thiol drugs (CBTD)
 - Agents most commonly used include α -mercaptopropionyl glycine (tiopronin) and D-penicillamine.
 - Thiol compounds contain sulfhydryl groups that undergo a disulfide exchange reaction with cysteine to produce two molecules of cysteine bound to the CBTD, a complex that is 50 times more soluble than cystine.
 - Chelation therapy is usually necessary if 24-hour urine cystine concentration remains > 2,000 $\mu\text{mol/L}$.

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- When maximum tolerated doses of tiopronine, high fluids, and alkalinization fail, penicillamine is the next line of medication for cystinuria. It also forms a more soluble disulfide with cysteine.
 - ESWL addresses the existing stones but doesn't prevent new formation, which is the primary issue here

WINNER

- Dr Anju Sukumaran,
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congratulations

The image features a solid black background. At the top, there is a decorative border composed of several overlapping, wavy bands of color. From left to right, these bands include shades of yellow, orange, red, and a hint of green and blue on the far right. The text "Thank you" is centered in the black area.

Thank you