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# The Clinical Picture

## Purple urine in a woman with chronic kidney disease

A 70-YEAR-OLD WOMAN presents with 3 days of fever (temperatures up to 38°C), abdominal pain, and purple-colored urine (FIGURE 1). She has stage 4 chronic kidney disease secondary to diabetes and hypertension. Her diabetes is controlled with insulin, and her hypertension with irbesartan (Avapro) 150 mg daily. She also has neurogenic bladder, managed by a urinary catheter for the last 6 months.

On examination, her blood pressure is 138/70 mm Hg, respiratory rate 18 breaths/minute, and heart rate 80 beats/minute. Her abdomen is soft. She has never undergone abdominal surgery. She is not taking any medications that may have caused urine discoloration.

Plain radiography of the abdomen reveals no abnormal gas. Laboratory test results are as follows:

- White blood cell count  $15.1 \times 10^9/L$  (reference range 4–11), with 85% neutrophils (reference range 39.5%–74%)
- C-reactive protein 6.27 mg/dL (reference range 0.0–1.0)
- Blood urea nitrogen 54 mg/dL (reference range 8–25)
- Serum creatinine 2.5 mg/dL (0.70–1.40)
- Estimated glomerular filtration rate 20 mL/min/1.73 m<sup>2</sup> (< 60 is sufficient for the diagnosis of chronic kidney disease)
- Liver function tests are normal
- Urine pH 8.0 (4.80–7.80); urine is positive for nitrates and for marked pyuria and bacteriuria.



FIGURE 1. Purple urine in the urine bag and tubing.

Urine culture yields more than 100,000 colony-forming units of *Pseudomonas aeruginosa*, *Morganella morganii*, and *Proteus vulgaris*. These results and the patient's presentation point to a diagnosis of purple urine bag syndrome. After placement of a new urinary cath-

eter and 7 days of intravenous ciprofloxacin (Cipro) 250 mg every 12 hours, the color of her urine returns to normal.

### ■ PURPLE URINE BAG SYNDROME

Purple urine bag syndrome is rare, and catheter-associated urinary tract infection is the main cause.<sup>1</sup> However, it has also been associated with intestinal intussusception.<sup>2</sup> In our patient, the examination and radiography ruled out intussusception.

Factors reported to be involved in the development of this syndrome include older age, female sex, chronic constipation, chronic urinary catheterization, alkaline (common) or acidic (uncommon) urine, and a higher bacterial load in the urine.<sup>3,4</sup>

The pathogenesis of purple-colored urine<sup>4,5</sup> is thought to start with the metabolism of dietary tryptophan by intestinal bacteria to indole. Indole is then absorbed into the portal circulation and is converted to indoxyl sulfate, which is excreted into the urine. In vitro experiments<sup>4,5</sup> have shown that certain bacteria in the urine produce indoxyl sulfatase and indoxyl phosphatase, which break down indoxyl sulfate to indoxyl. Indoxyl can then be converted to indigo or indirubin in alkaline<sup>5</sup> or acidic<sup>4</sup> urine. When blue indigo and red indirubin mix together, the result is purple.<sup>4,5</sup>

### ■ REFERENCES

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Bacteria that possess indoxyl sulfatase or indoxyl phosphatase include *P aeruginosa*, *M morganii*, *P vulgaris*, *Escherichia coli*, and *Providencia* species.<sup>5,6</sup> However, not all bacteria of the same species produce the enzymes required for the formation of purple urine.<sup>5</sup> This may explain the rarity of this syndrome despite the common occurrence of urinary tract infection in patients with risk factors for purple urine bag syndrome.

### ■ CHRONIC KIDNEY DISEASE: A POTENTIAL RISK FACTOR

Chronic kidney disease was shown to be a risk factor for purple urine bag syndrome in a small cohort study of Taiwanese patients.<sup>7</sup> The serum and urine levels of indoxyl sulfate increased markedly in patients who had chronic kidney disease or who were undergoing dialysis because of impaired renal clearance.<sup>6</sup> Furthermore, indoxyl sulfate, which plays an important role in this syndrome, is also cytotoxic and may increase the rate of renal failure in uremic rats.<sup>4</sup>

Although purple urine itself is usually considered benign,<sup>3</sup> it should prompt an evaluation for urinary tract infection, especially in patients with kidney disease. Failure to treat the underlying infection can lead to septicemia or Fourier gangrene.<sup>1,3</sup> ■

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