Medicine in Pictures: Purple Urine Bag Syndrome

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An 86-year-old lady was admitted from her residential home with acute on chronic confusion, new symptoms of expressive and receptive dysphasia, dysphagia, vacant episodes and urinary incontinence. She had a previous significant history of haemorrhagic stroke with residual right sided weakness, atrial fibrillation, hypertension, and moderate dementia. Following a CT head, this lady was started on acyclovir for encephalitis. She failed to respond to treatment, and developed constipation. With careful consideration of her poor prognosis and quality of life, this lady was placed on the End of Life Pathway. She was catheterised for comfort. Nine days after initial insertion of the urinary catheter, purple urine was noted in the catheter bag with yellow urine in the tubing leading to the bag. Urine dipstick showed Blood ++, Protein ++, Leuc +, Nit -ve, Glu ve, Ketone +, pH 8.0. Urine microscopy showed: WCC 454, RBC 279, epithelial cells 52, no casts. Urine culture revealed heavy mixed growth with multiple organisms.



Question:

What is the diagnosis?

Answers:

- 1. Porphyria
- 2. Propofol infusion syndrome
- 3. Purple urine bag syndrome
- 4. Blue diaper syndrome

Differential diagnoses:

Discoloration of urine can be caused by trauma if blood stained, urinary tract infections, ingestion of dye (methylene blue), medications (amitriptyline, indomethacin, triamterene, flutamide, and phenol).

Explanation:

Porphyria usually presents with severe pain with neuropsychological symptoms or photosensitivity, and urine discoloration is likely to occur from initial onset of disease.

Propofol is an anaesthetic agent, excreted in the urine as phenol derivatives which can cause a green urine discolouration¹. This medication is unlicensed for End of Life Pathway. Propofol infusion syndrome is associated with prolonged high dose infusion, but is not always accompanied by urine discoloration.

Blue diaper syndrome is an inherited metabolic disorder of tryptophan with presentation at infancy²⁻³.

Correct answer

Purple urine bag syndrome (PUBS)

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PUBS is an uncommon condition with purple discoloration of the urine catheter system. This phenomenon is due to the presence of indigo and indirubin in the collected urine. PUBS was first published in 1978⁴. Some academics would argue that PUBS was reported even earlier historically as an observation in Sir Henry Halford's bulletin in 1811⁵⁻⁶. Two recent literature reviews suggested the prevalence of PUBS is as high as 9.8% in institutionalized patients with long-term urinary catheterisation^{8-9, 12}.

A triad of key factors are suggested as cause of PUBS:

- High level of tryptophan in the gut due to diet intake or bowel stasis
- 2. Long term catheterisation⁸
- Urinary tract infection (uti) with bacteria possessing indoxyl phosphatase and sulphatase enzymes, commonlyprovidencia stuarttiand rettgeri, pseudomonas

auruginosa, proteus mirabilis, escherichia coli, klebsiella pneumoniae,morganella, citrobacter species, group b *streptococci* and *enterococci*^{8, 13}.

It is understood that bowel stasis causes accumulation of tryptophan, which leads to an increase in urinary indoxyl sulphate (UIS). In the presence of indoxyl phosphatase and sulphatase enzyme activities, whilst collected in the catheter system, UIS is degraded to form a mixture of indigo and dissolved indirubin in the plastic¹¹, coating the catheter system with a purple appearance. Intensity of discoloration is deeper the longer the urine is in contact with the catheter plastic^{7, 10-12}. The urine does not appear purple prior to entering the catheter.

Recent literature⁷⁻⁸ also suggested female gender, alkaline urine, bed bound debilitated patient population, PVC material⁷ and institutionalization are further predisposing factors of PUBS.

Management of PUBS requires catheter change and treatment of underlying UTI.

Good catheter hygiene and shorter duration of catheterisation can reduce PUBS¹.

Competing Interests None declared

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REFERENCES

- Foot C, Fraser JF. Uroscopic Rainbow: Modern Matula Medicine. Postgrad Med J 2006;82(964):126–129.
- Drummond K, Miachael A, Ulstrom R, et al. The Blue Diaper Syndrome: Familial Hypercalcemia with Nephrocalcinosis and Indicanuria; A new familial disease, with definition of the metabolic abnormality. Am J Med. 1964 Dec;37:928-48.
- Cochat P, Pichault V, Bacchetta J, et al. NephrolithiasisRelated to Inborn Metabolic Diseases. Pediatr Nephrol 2010;25(3):415–424.
- 4. Barlow G, Dickson J. Purple Urine Bags. Lancet 1978;1:220-221.
- Arnold W. King George III's Urine and Indigo Blue. Lancet 1996;347:1811-1813.
- Macalpine I, Hunter R. The "Insanity" of King George III: A Classic Case of Porphyria. BMJ 1966;1:65-71.
- Su F, Chung S, Chen MH, et al. Case Analysis of Purple Urine-Bag Syndrome at a Long-Term Care Service in a Community Hospital. Chang Gung Med J 2005;28:636-642.
- Khan F, Chaudhry M, Qureshi N, et al. PurpleUrine Bag Syndrome: An Alarming Hue? A Brief Review of the Literature. Int J Nephrol. 2011;2011:419213.
- Dealler S, Belfield P, Bedford M, et al. Purple Urine Bags. J Urol1989;142(3):769–770.
- Al-Sardar H, Haroon D, Purple urinary bag syndrome. Am J Med 2009;122(10):e1-2.
- Dealler S, Hawkey P, Millar M. Enzymatic Degradation of Urinary Indoxyl Sulfate by Providencia Stuartii and Klebsiella Pneumoniae Causes the Purple Urine Bag Syndrome. J Clin Microbiol 1988;26:2152-2156.
- Lin C, Huang H, Chien C, et al. Purple Urine Bag Syndrome in Nursing Homes: Ten Elderly Case Reports and a Literature Review. Clin Interv Aging 2008;3(4):729-734.
- Vallego-Manzur F, Mireles-Cabodevila E, Varon J. Purple Urine Bag Syndrome. Am J Emerg Med 2005;23:521-524.