

Maculopathy caused by pentosan polysulfate

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1 Pentosan polysulfate (PPS) is a mainstay for treatment of bladder pain associated with interstitial cystitis

Interstitial cystitis is a chronic bladder disorder causing pelvic pain and urinary frequency or urgency, with an estimated prevalence of 2.7%–6.5% among adult women.¹ Pentosan polysulfate may buffer the bladder epithelium against irritants and is the only oral medication approved by Health Canada for treatment of interstitial cystitis.¹ It is typically administered orally at a dose of 100 mg 3 times daily.

2 Maculopathy is associated with longer duration of PPS use

A large retrospective study noted an increased likelihood of macular disease after 7 years of cumulative PPS exposure (odds ratio 1.41, 95% confidence interval 1.09–1.83).² A cross-sectional study of 50 patients with history of PPS therapy found that 15%–20% exhibited evidence of macular toxicity with formal retinal imaging. The prevalence of macular disease increased to 50% among patients with cumulative doses exceeding 1500 g.³

3 Maculopathy caused by PPS may masquerade as age-related macular degeneration

Maculopathy caused by PPS is associated with difficulty reading and poor vision in low-light conditions. Patients with PPS maculopathy are often given diagnoses of more common conditions, such as age-related macular degeneration.²

4 Macular disease may progress even after cessation of PPS

The proposed pathogenesis of PPS maculopathy involves disruption of the extracellular matrix surrounding the photoreceptors or direct toxicity to the retinal pigment epithelium.^{2–4} It remains unclear whether disease trajectory is altered by drug cessation.⁴ Case series have documented progressive maculopathy for up to 6 years following discontinuation of PPS.⁵

5 Patients exposed to PPS who report disturbed vision should undergo ophthalmic screening

Prescribers of PPS and primary care providers should be aware that the Canadian Urological association suggests such screening.⁵ More recent ocular studies have recommend baseline retinal examination and imaging, especially in patients with cumulative PPS exposure greater than 500 g, but more evidence is required to further refine screening guidelines.^{3,5}

References

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