

**Low availability of haematin (hemin) in Pakistan**Fatima Ahsan<sup>1</sup>, Muhammed Iffham Imtiyas<sup>2</sup>, Abdul Raheem Rizvie<sup>3</sup>

*Dear Madam,* Acute Intermittent Porphyria (AIP) is an autosomal dominant disorder that results from a defect in the enzyme named porphobilinogen deaminase.<sup>1</sup> It is symptomatic porphyria, involving the accumulation of porphyrins and porphyrin precursors due to impaired conversion in haeme synthesis. Acute Intermittent Porphyria is clinically characterized by acute episodes of multiple gastrointestinal and neurologic symptoms; during which the patient remains healthy between episodes.<sup>2</sup> Acute Intermittent Porphyria episodes are commonly observed in post pubertal females. The most common symptoms include periodic abdominal pain, vomiting and hypertension followed by neurologic damage that leads to peripheral and autonomic neuropathies (mostly motor) and psychiatric manifestations.<sup>2</sup> Hormonal cycle regulation may worsen porphyria, since its symptoms coincide with the menstrual cycle. Although positive response has been noted in hormonal suppression by gonadotropin releasing hormone agonists.<sup>3</sup>

Currently, the most widely used treatment is hemin, a form of haeme that can be given intravenously. Its administration reduces the haeme deficit, hence limits the production of porphyrins precursors by the body. Availability of hemin in Pakistan is not promising while alternatives such as intravenous infusion of glucose, and pain relievers like Nalbuphine, a man-made opioid analgesic, and Morphine are short-term treatments. Haeme Arginate (HA), a compound of haeme and arginine, is equivalent to hematin in the treatment of AIP. Long-term weekly intravenous infusions of prophylactic HA prevent severity and frequency of porphyric attacks in patients with AIP.<sup>4</sup> While haeme therapy is recommended if a patient exhibits neurological impairment, liver transplant appears to be the last resort in patients with recurrent clinical

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manifestations.

To effectively mediate the growing need of Hemin in Pakistan, a systematic plan with effective and immediate steps must be introduced. The regulatory authorities must expedite the approval and licensing process of appropriate medication that would meet the growing need of the country, while not compromising on its quality. The possibility of local production should be explored, while strengthening international ties aimed at importing affordable medication. The government should focus on initiating partnerships with international organizations, non-governmental organizations (NGOs) and global health initiatives. Nationwide workshops and informative campaigns towards healthcare work should be a priority, to raise awareness of the situation and the proper steps that should be taken to minimize its adverse effects.

Empowering patient advocacy groups would be beneficial in recognizing any shortcomings of the plan and to divert resources accordingly.

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