Prescribing Restrictions in Non-Acute Cutaneous Porphyria

Summary

The cutaneous porphyrias (in which acute attacks of porphyria do not occur) include:

- porphyria cutanea tarda
- erythropoietic protoporphyria
- x-linked erythropoietic protoporphyria
- congenital erythropoietic porphyria

In these conditions, bullous skin lesions or acute painful photosensitivity occur due to the accumulation of porphyrins in the skin.

"Safe drug lists" are not relevant and do not need to be adhered to in patients with cutaneous porphyrias.

Congenital Erythropoietic Porphyria (CEP)

CEP (also known as Günther's disease) is an exceptionally rare and severe inherited disease with a wide variety of features that occur due to decreased activity of the enzyme uroporphyrinogen III synthase.

Treatment modalities that stimulate erythropoiesis (such as iron supplements and erythropoietin) should generally be avoided. CEP related anaemia is usually treated with transfusion. Always discuss with a porphyria expert centre.

New-borns diagnosed with CEP and that have hyperbilirubinemia should not have photodynamic therapy as this will worsen skin symptoms.

Erythropoietic Protoporphyria (EPP)

EPP is a rare form of cutaneous porphyria. It causes painful acute photosensitivity usually within seconds to minutes after sunlight or very bright indoor light exposure.

Due to the risk of serious cholestatic liver disease which occur in a small minority of patients, sensible intake of alcohol, maintaining a healthy weight and Hep A and B vaccination is now recommended by many porphyria centres.

Many EPP patients have a mild microcytic anaemia as part of the EPP phenotype. Iron supplementation should be avoided in EPP unless a clear iron deficiency is demonstrated or the anaemia is severe and symptomatic. If oral iron is required, it should be given very cautiously at a low dose and liver functions should be monitored appropriately. Discuss with a porphyria expert centre.

X-linked Erythropoietic Protoporphyria (XLEPP)

XLEPP clinically presents in a similar manner to EPP but is caused by altered activity of a different enzyme in the haem biosynthesis pathway. Therefore, similar advice is given and no prescribing restrictions are required.

In contrast, however, anecdotal evidence suggests that iron supplementation may aid symptoms - compared to making them worse for EPP. Further information can be found on the International Porphyria Network website.

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Porphyria Cutanea Tarda (PCT)

PCT is the most common type of porphyria and presents with bullous skin lesions due to a deficiency in the enzyme uroporphyrinogen decarboxylase. There are two types of PCT:

- Acquired (sporadic) form which is the most common and usually associated with liver disease
- The less common familial form

Treatment is often very successful and results in remission of the condition, although PCT may relapse. When PCT is in remission, there are no prescribing restrictions.

For both types of PCT it is recommended that alcohol consumption is stopped as this is an associated, causative risk factor.

Active PCT

PCT is classed as active when patients have at least one of the following:

- active skin lesions
- increased concentrations of porphyrins in the urine or blood

There are a number of drugs that should be avoided when PCT is active as they may increase porphyrin concentrations or be involved as a causative factor.

Caution should be exercised when using:

- **Oestrogen**: oestrogen has been shown to have an impact on PCT expression and has been indicated as a causative factor. This should be especially noted for people on hormone treatments; who are taking an oral contraceptive; or those undergoing cancer treatments.

- **Iron supplements**: a hepatic iron rich environment is central to the pathogenesis of PCT so caution should be exercised if iron supplements are required, and the condition monitored if supplementation is essential.

- **High dose hydroxychloroquine or chloroquine:** low doses of hydroxychloroquine and chloroquine should be given if used as a treatment e.g. 200mg hydroxychloroquine twice a week or 125mg chloroquine twice a week. These low doses promote porphyrin excretion. Normal doses can cause a serious adverse reaction in active PCT, with pyrexia and acute hepatitis, and should be avoided.

  If medications are to be used for malaria prevention, seek advice from a porphyria expert centre.

Previously it was also suggested that methotrexate was contraindicated, however, the latest information suggests that it is now safe to use with PCT.

Mae'r daflen hon hefyd ar gael yn Gymraeg. This leaflet is also available in Welsh.