

THINK PNH:

A SPOTTERS GUIDE FOR ANTI-COAGULATION HCPs

Paroxysmal nocturnal haemoglobinuria (PNH) is a rare blood condition which leaves patients vulnerable to thrombotic events.

It is caused by a bone marrow stem cell mutation which produces red blood cells that are missing some of their cell surface shields. This makes them vulnerable to destruction by a part of the immune system called complement and can cause anaemia. However as their bone marrow may work hard to compensate for the loss, their haemoglobin may actually be within normal range which can make it hard to spot. In many patients other blood cells such as platelets or white blood cells can be lower than the normal range (cytopenia).

This process of red cell destruction, 'haemolysis', may also cause other problems that may not at first seem related to a blood condition. For this reason, it is difficult to diagnose PNH and patients can often suffer harm before the underlying cause of their problems is understood.

These symptoms may include any of the following. Each patient is different and symptoms may be transient:

| | | | |
|----------------------|------------|-----------------|---------------------|
| Thrombosis | Dark Urine | Abdominal pains | Shortness of breath |
| Erectile dysfunction | | Dysphagia | Fatigue |

If you have a patient who has a thrombosis without a clear cause **THINK PNH and request an LDH and reticulocyte count especially if they are:**

- **Young**
- **Have, or have ever had any cytopenias (low red cells, white cells or platelets)**
- **Any other organ dysfunction**

If the LDH or Reticulocytes are raised you should request a PNH screen.

If the screen is positive, they will require urgent referral to the PNH Service.

In the setting of PNH one blood clot predicts another so it is important that the referral is made promptly.

LONDON PNH SERVICE, KING'S COLLEGE HOSPITAL kch-tr.kingspnh@nhs.net 0203 299 1039 / 3520 / 1153

LEEDS, ST. JAMES HOSPITAL pnh.leeds@nhs.net Medical emergency direct on call service (17:00 – 08:30 hours): 07920 535918