Barts Health NHS Trust

Haemoglobinopathy Coordinating Centre

Sickle Cell in Primary Care

The acute sickle cell illness pathway was introduced to facilitate the rapid evaluation of children with sickle cell disease who present to primary care with an acute illness. These patients have an increased vulnerability to infections, particularly to invasive pneumococcal disease and this is a significant cause of morbidity and mortality, especially in children under 5 years old.

Early identification and assessment of acute complications in a primary care setting are crucial, with prompt referral to a specialist centre when necessary.

Use of the pathway in a primary care setting

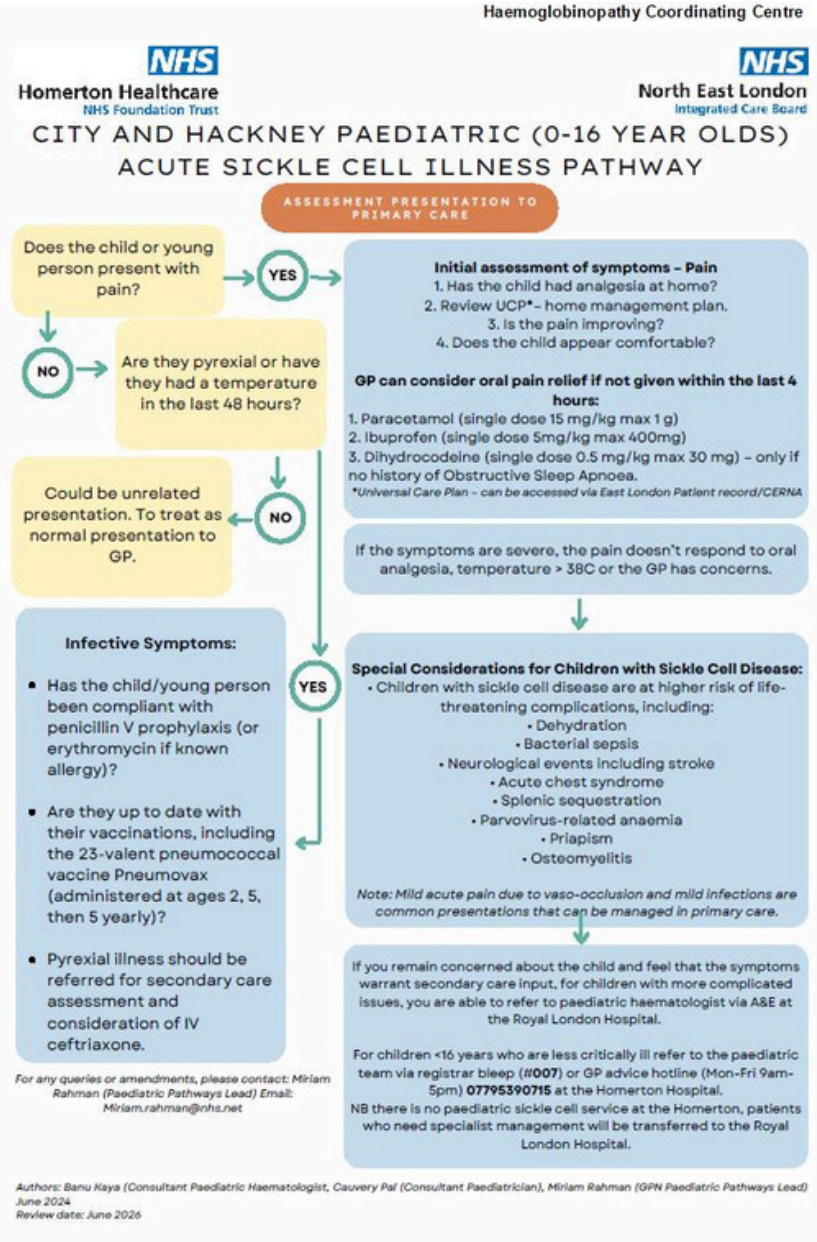
The pathway was developed in response to a patient death where sepsis was not recognised despite multiple presentations to the GP. It provides a flowchart for paediatric patients who present to the GP with pain and/or fever, to aid triage and escalation in conjunction with their Universal Care Plan (UCP). Following on from a number of Serious Incidents (SIs), we are reminding primary care centres of the importance of accurately assessing and triaging these patients, ensuring early intervention and referral to secondary and tertiary care when necessary. If the pathway is used alongside the UCP, paediatric patients will receive timely support, helping to prevent potential mortality and morbidity.

It is also important to consider regular reviews of the patients' medication including compliance, vaccination history and any updates made to their UCP.

Examples of how to use the pathway

3 year old with homozygous sickle cell disease. Parents bring her to the GP surgery as she has been experiencing abdominal discomfort, reduced food and drink intake and has had a fever overnight. Parents report that she is miserable and is not her normal self. She is up to date with her vaccinations. She takes folic acid and Pen V however, her compliance is suboptimal. Considering; 1. No history of pain. 2. A history of pyrexia in the last 48 hours. 3. Patient has not been compliant with their prophylactic antibiotic. **Patient's UCP should be reviewed and refer patient to the named secondary centre for assessment.**

15 year old girl presents to the GP with a 3 day history of fever (up to 40C), cough, aches, nausea and vomiting. She has had progressive chest, back and abdominal pain over the last few hours. She has a past medical history of splenectomy. She takes folic acid, Pen V and hydroxycarbamide with no issues. Prior to presentation to the GP, she has had paracetamol, ibuprofen and dihydrocodeine. On assessment, she appears unwell, with tachycardia, pyrexia and reduced air entry on both sides. Considering; 1. Presenting with pain AND pyrexia in the last 48 hours. 2. She has not responded to paracetamol, ibuprofen AND dihydrocodeine. **Patient's UCP should be reviewed and urgently refer patient to named secondary centre for immediate medical attention. Giving the history of the patient, an ambulance should be called and patient should be blue lighted to the nearest A&E.**



Key points

- Red flags: **Severe pain, pyrexia in the last 48 hours**
- Risk factors: **Non-compliance to antibiotic prophylaxis, vaccinations not up to date (particularly pneumovax), pyrexial illness**
- Special considerations: **Dehydration, bacterial sepsis, neurological events, acute chest syndrome, splenic sequestration, parvovirus related anaemia, priapism, osteomyelitis.**

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