Yorkshire and Humber Children and Young People’s Cancer Network

Referral, Diagnosis & Staging Protocol
### Document Control

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<tr>
<th>Title</th>
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<tr>
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<td>Dr S Picton and Ms R Hollis</td>
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<td>Yorkshire and Humber Children’s &amp; Young People Cancer Network</td>
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### Version Control

<table>
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<td>1.0</td>
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### Contributors to current version

<table>
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<tr>
<th>Contributor</th>
<th>Author/Editor</th>
<th>Section/Contribution</th>
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<tr>
<td>CYP Group</td>
<td></td>
<td>All</td>
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## Information Reader Box

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<thead>
<tr>
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<th>Referral, Diagnosis and Staging Protocol</th>
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<tr>
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</tr>
</tbody>
</table>
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LS9 7TF  
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Table of Contents

I DOCUMENT CONTROL .................................................................................................................. 2
II INFORMATION READER BOX ....................................................................................................... 3
III TABLE OF CONTENTS ................................................................................................................. 4

1 REFERRAL DIAGNOSIS AND STAGING PROTOCOL ................................................................. 5
  1.1 REFERRAL FROM PRIMARY CARE ....................................................................................... 5
  1.2 FIRST SEEN AT POSCU/DGH ............................................................................................ 5
    1.2.1 Suggested Investigations at POSCU/DGH ................................................................. 5
  1.3 LOCAL MANAGEMENT OF PATIENTS WHO PRESENT AS ONCOLOGICAL EMERGENCIES ............................................................ 6

2 APPENDIX 1 - REFERRAL GUIDELINES FOR SUSPECTED CANCER IN CHILDREN .... 10
  2.1 GENERAL RECOMMENDATIONS: ....................................................................................... 10
  2.2 SPECIFIC RECOMMENDATIONS: ....................................................................................... 11
    2.2.1 Leukaemia (children of all ages) .................................................................................. 11
    2.2.2 Lymphomas: ............................................................................................................... 11
    2.2.3 Brain and CNS Tumours: ............................................................................................ 12
    2.2.4 Neuroblastoma (all ages) ........................................................................................... 13
    2.2.5 Wilms’ tumour (all ages) ............................................................................................ 13
    2.2.6 Soft tissue sarcoma (all ages) ..................................................................................... 13
    2.2.7 Bone sarcomas (osteosarcoma and Ewing’s sarcoma) (all ages) ................................. 14
    2.2.8 Retinoblastoma (mostly children aged under 2 years) .............................................. 14
  2.3 INVESTIGATIONS .................................................................................................................... 15
1 Referral Diagnosis and Staging Protocol

This protocol describes the referral pathway for Children and Young People (CYP) from 0 - 15 years (up to their 16th birthday) who are suspected of having cancer. It is written to support the high level Children’s Cancer (0 -16) pathways, and the pathway for Children with Brain and CNS Tumours. http://www.ycn.nhs.uk/html/pathways/clinical/children.php

This protocol provides guidance on the referral routes from primary and community care, to secondary care and on to the tertiary speciality ‘Principal Treatment Centre (PTC) in Leeds. Referral from secondary care to the PTC should through direct Consultant to Consultant telephone contact, along with a written referral. (Table on page 8 sets out contact numbers for both secondary care and PTC clinicians).

1.1 Referral from primary care

Primary Care Practitioners should establish whether the child’s signs and symptoms fulfil the criteria for urgent referral for suspected cancer as defined in the NICE 2005 Referral Guidelines [see Appendix 1 and http://guidance.nice.org.uk/CSGCYP ].

Initial referral should be to the named Lead Paediatrician (or on-call Paediatrician if out of hours) in the local secondary care Paediatric Oncology Shared Care Unit (POSCU). If the local DGH does not provide a Shared Care service (see table page 8) then referral should be made to the on-call Paediatrician at the local DGH which provides children’s services.

If an urgent referral is made via the Referral Management System (via 2 week cancer wait system) this should be followed up by direct contact with the local service.

If a blood test carried out in Primary Care is reported by the Haematological Malignancy Diagnostic Service (HMDS) to indicate Leukaemia, patients may be referred directly to the Principal Treatment Centre in Leeds. If imaging investigations are carried out in Primary Care and reported as highly suspicious of malignancy, patients may be referred directly to the Principal Treatment Centre in Leeds. In these cases, notification of referral should be made to local secondary paediatric service (POSCU or non-POSCU DGH).

1.2 First seen at POSCU/DGH

In children presenting with signs and symptoms which may be due to malignant disease (or relapse) the local hospital will undertake those haematological investigations, biochemical investigations and imaging studies (ultrasound, CT and MRI - if available) which are necessary to exclude the diagnosis of malignant disease.

1.2.1 Suggested Investigations at POSCU/DGH

(Further information can be found in the NICE 2005 Referral Guidelines - see Appendix 1)

- Blood tests
  - Full Blood Count, Film and Blood specimen to HMDS if leukaemia suspected
  - U&E (including urate)
  - LFT

***VALID ON DATE OF PRINTING ONLY - all guidelines available at http://www.ycn.nhs.uk/ ***
• LDH if Lymphoma suspected
• AFP and Beta HCG if liver tumour or other solid tumour suspected

Radiology

• Chest X Ray if Leukaemia/Lymphoma suspected
• Plain X Ray of affected area if bone tumour suspected
• USS for abdominal/neck masses
• Other imaging such as CT scan and MRI as indicated; this may usefully be discussed with on-call consultant at PTC

Other

• Urine for Catecholamines if neuroblastoma suspected

Diagnostic and staging investigations which should only be performed at a PTC include:

• bone marrow, lumbar puncture or paracentesis for cytology;

• surgical biopsy [except for excision biopsy of superficial lymph nodes which may take place in a POSCU with appropriate surgical expertise and pathology infrastructure:] Cases can be discussed with the paediatric surgical or oncology team at the PTC but if a tumour is likely biopsy should take place in Leeds as fresh tissue is required for cytogenetics;

• detailed staging investigations, such as Bone Scan or MiBG

If initial investigations indicate that malignant disease is likely the patient should be referred immediately to the Principal Treatment Centre in Leeds.

The Information Pathway for Parents of Children and Young People with Cancer (http://www.ycn.nhs.uk/html/pathways/info/childrens.php) supports the initial steps in the children’s pathway through the process of referral, diagnostic procedures and tests, diagnosis, and staging and treatment.

1.3 Local Management of patients who present as oncological emergencies

The following situations are oncological emergencies and should be discussed with the consultant on-call at the PTC in Leeds:

• High white cell counts - white cell counts over 100x10^9/l. Blood should be sent directly to HMDS in Leeds as, in the presence of a high count, the diagnosis of leukaemia can be established using blood rather than bone marrow. Hyperhydration and Allopurinol treatment should be commenced prior to urgent transfer to Leeds.

• Superior vena cava/superior mediastinal syndrome - patient should be assessed in the local hospital to determine whether urgent ambulance transport to Leeds would be safe or whether Paediatric Intensive Care retrieval is necessary.
• Acute raised intra-cranial pressure – such patients should be investigated urgently, and consultation undertaken with the Paediatric Neurosurgeon On-Call in Leeds (see Guideline for the management of Children and Young People with Intracranial pressure). http://www.ycn.nhs.uk/html/publications/guidelines_paediatric.php
Immediate management may require the involvement of Paediatric Intensive Care

• Acute spinal cord compression – if strongly suspected, the patient should be discussed with the on-call paediatric oncologist, who may also advise or undertake discussion with the on-call neurosurgeon covering paediatric neurosurgery and on-call clinical oncology team in Leeds. (See Guideline for suspected spinal cord compression in children and young people < 18 years old) http://www.ycn.nhs.uk/html/publications/guidelines_paediatric.php
A plan for urgent investigation and subsequent management will be jointly decided.

• Septic shock - the patient should be stabilised in the local hospital and, when deemed fit for travel, transferred urgently to Leeds (see YHCYPCN Guideline http://www.ycn.nhs.uk/html/publications/guidelines_paediatric.php for Referral of patients with Febrile Neutropenia).
The table below details the contact points of the PTC in Leeds, and of local hospitals which refer to the PTC.

<table>
<thead>
<tr>
<th>CCG - Clinical Commissioning Group</th>
<th>POSCU or local DGH which provides children’s services</th>
<th>Contact point in POSCU and/or DGH</th>
</tr>
</thead>
<tbody>
<tr>
<td>Airedale, Wharfedale &amp; Craven</td>
<td>POSCU</td>
<td>Dr Analies Mawdsley or on-call Paediatrician = contacted via Airedale Hospital switchboard</td>
</tr>
<tr>
<td>Harrogate &amp; Rural District</td>
<td>Airedale NHS Foundation Trust</td>
<td>Tel: (01535) 652511 ask to page on-call.</td>
</tr>
<tr>
<td>Airedale, Wharfedale &amp; Craven</td>
<td>Bradford Teaching Hospital NHS Foundation Trust</td>
<td>On call consultant paediatrician to be contacted via switchboard</td>
</tr>
<tr>
<td>Bradford District</td>
<td></td>
<td>Tel: (01274) 542200</td>
</tr>
<tr>
<td>Bradford City</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Calderdale</td>
<td>POSCU</td>
<td>Dr Gill Sharpe Consultant Paediatrician via switchboard or on-call Paediatrician contacted via Calderdale Royal Hospital Switchboard</td>
</tr>
<tr>
<td>Greater Huddersfield</td>
<td>Calderdale and Huddersfield NHS Foundation Trust</td>
<td>Tel: (01422) 357171</td>
</tr>
<tr>
<td>North Kirklees</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Leeds North</td>
<td>PTC</td>
<td>On-call Paediatric Oncologist or Haematologist contacted via Leeds General Infirmary switchboard</td>
</tr>
<tr>
<td>Leeds West</td>
<td>Leeds Teaching Hospital NHS Trust</td>
<td>Tel: (0113) 2432799</td>
</tr>
<tr>
<td>Leeds South &amp; East</td>
<td></td>
<td>Fax (0113) 2470248</td>
</tr>
<tr>
<td>Harrogate &amp; Rural District</td>
<td>Harrogate and District Hospitals NHS Foundation Trust</td>
<td>On-call Paediatrician via Switchboard</td>
</tr>
<tr>
<td></td>
<td></td>
<td>(01423) 885959</td>
</tr>
<tr>
<td>Wakefield</td>
<td>Mid Yorkshire Hospital NHS Trust</td>
<td>On-call Paediatrician</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Tel: (01924) 512050</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Fax: (01924) 816175</td>
</tr>
<tr>
<td>North Kirklees</td>
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</table>
Principal Treatment Centre - LEEDS

Address: Paediatric Oncology and Haematology Admin Offices
D Floor Martin Wing
Leeds General Infirmary
Great George Street
Leeds
LS1 3EX

Contacts Details: On-call Consultant Paediatric Oncologist and Haematologist contacted via Main LGI Switchboard
Tel: (0113) 2432799

Admin Office: Tel: (0113) 3928191 Fax: (0113) 3928488

Paediatric Wards: Ward L31 (0113) 3927431
Ward L32 BMT (0113) 3927432
Ward L33 Teenage & Young Adult (0113) 3927433
Children’s Haematology & Oncology Day Unit (0113) 3927179

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<thead>
<tr>
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<tr>
<td>Vale of York</td>
<td>POSCU York Teaching Hospital NHS Foundation Trust</td>
<td>Dr Jill Mant or on-call Consultant Paediatrician via switchboard Tel: (01904) 725314</td>
</tr>
<tr>
<td>Yorkshire</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Scarborough &amp; Ryedale</td>
<td>POSCU Scarborough Hospital</td>
<td>Dr Jill Mant or on-call Consultant Paediatrician on call Phone (01723) 368111 Fax (01723) 385282</td>
</tr>
<tr>
<td>East Riding of Yorkshire</td>
<td></td>
<td></td>
</tr>
<tr>
<td>NHS Wakefield District</td>
<td>POSCU Calderdale and Huddersfield NHS Foundation Trust</td>
<td>Dr Gill Sharpe Consultant Paediatrician or on-call Paediatrician contacted via Calderdale Royal Hospital Switchboard Tel: (01422) 357171 On-Call Consultant Paediatrician Tel: (01924) 512050 Fax: (01924) 816175</td>
</tr>
<tr>
<td>Hull</td>
<td>POSCU Hull and East Yorkshire Hospitals NHS Trust</td>
<td>Dr Ashwini Kotwal or on-call Consultant Paediatrician Hull Royal Infirmary Tel: (01482) 674039</td>
</tr>
<tr>
<td>East Riding of Yorkshire</td>
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Appendix 1 - Referral guidelines for suspected cancer in children

2.1 General recommendations:

Children and young people who present with symptoms and signs of cancer should be referred to a paediatrician or a specialist children's cancer service, if appropriate.

Childhood cancer is rare and may present initially with symptoms and signs associated with common conditions. Therefore, in the case of a child or young person presenting several times (for example, three or more times) with the same problem, but with no clear diagnosis, urgent referral should be made.

The parent is usually the best observer of the child’s or young person’s symptoms. The primary healthcare professional should take note of parental insight and knowledge when considering urgent referral.

Persistent parental anxiety should be a sufficient reason for referral of a child or young person, even when the primary healthcare professional considers that the symptoms are most likely to have a benign cause.

Persistent back pain in a child or young person can be a symptom of cancer and is indication for an examination, investigation with a full blood count and blood film, and consideration of referral.

There are associations between Down’s syndrome and leukaemia, between neurofibromatosis and CNS tumours, and between other rare syndromes and some cancers. The primary healthcare professional should be alert to the potential significance of unexplained symptoms in children or young people with such syndromes.

The primary healthcare professional should convey information to the parents and child/young person about the reason for referral (NICE Guideline – Referral for suspected cancer) and which service the child/young person is being referred to so that they know what to do and what will happen next.

The primary healthcare professional should establish good communication with the parents and child/young person in order to develop the supportive relationship that will be required during the further management if the child/young person is found to have cancer.
2.2 Specific recommendations:

2.2.1 Leukaemia (children of all ages)
Leukaemia usually presents with a relatively short history of weeks rather than months. The presence of one or more of the following symptoms and signs requires investigation with full blood count and blood film:

- pallor
- fatigue
- unexplained irritability
- unexplained fever
- persistent or recurrent upper respiratory tract infections
- generalised lymphadenopathy
- persistent or unexplained bone pain
- unexplained bruising

If the blood film or full blood count indicates leukaemia then an urgent referral should be made.

The presence of either of the following signs in a child or young person requires immediate referral:

- unexplained petechiae
- hepatosplenomegaly

2.2.2 Lymphomas:
Hodgkin’s lymphoma presents typically with non-tender cervical and/or supraclavicular lymphadenopathy. Lymphadenopathy can also present at other sites. The natural history is long (months). Only a minority of patients have systemic symptoms (itching, night sweats, fever). Non-Hodgkin’s lymphoma typically shows a more rapid progression of symptoms, and may present with lymphadenopathy, breathlessness, superior vena-caval obstruction or abdominal distension.

Lymphadenopathy is more frequently benign in younger children but urgent referral is advised if one or more of the following characteristics are present, particularly if there is no evidence of local infection:

- lymph nodes are non-tender, firm or hard
- lymph nodes are greater than 2 cm in size
- lymph nodes are progressively enlarging
- other features of general ill-health, fever or weight loss
- the axillary nodes are involved (in the absence of local infection or dermatitis)
- the supraclavicular nodes are involved.

The presence of hepatosplenomegaly requires immediate referral.

Shortness of breath is a symptom that can indicate chest involvement but may be confused with other conditions such as asthma. Shortness of breath in association with the above signs, particularly if not responding to bronchodilators, is an indication for urgent referral.
A child or young person with a mediastinal or hilar mass on chest X-ray should be referred immediately.

### 2.2.3 Brain and CNS Tumours:

#### Children aged 2 years and older and young people

Persistent headache in a child or young person requires a neurological examination by the primary healthcare professional. An urgent referral should be made if the primary healthcare professional is unable to undertake an adequate examination. Headache and vomiting that cause early morning waking or occur on waking are classical signs of raised intracranial pressure, and an immediate referral should be made. The presence of any of the following neurological symptoms and signs should prompt urgent or immediate referral:

- new-onset seizures
- cranial nerve abnormalities
- visual disturbances
- gait abnormalities
- motor or sensory signs
- unexplained deteriorating school performance or developmental milestones
- unexplained behavioural and/or mood changes

A child or young person with a reduced level of consciousness requires emergency admission.

#### Children < 2 years

In children aged younger than 2 years, any of the following symptoms may suggest a CNS tumour, and referral (as indicated below) is required:

- **Immediate referral:**
  - new-onset seizures
  - bulging fontanelle
  - extensor attacks
  - persistent vomiting

- **Urgent referral:**
  - abnormal increase in head size
  - arrest or regression of motor development
  - altered behaviour
  - abnormal eye movements
  - lack of visual following
  - poor feeding/failure to thrive.

- **Urgency contingent on other factors:**
  - squint.
2.2.4 Neuroblastoma (all ages)

Most children and young people with neuroblastoma have symptoms of metastatic disease which may be general in nature (malaise, pallor, bone pain, irritability, fever or respiratory symptoms), and may resemble those of acute leukaemia. The presence of any of the following symptoms and signs requires investigation with a full blood count:

- persistent or unexplained bone pain (and X-ray)
- pallor
- fatigue
- unexplained irritability
- unexplained fever
- persistent or recurrent upper respiratory tract infections
- generalised lymphadenopathy
- unexplained bruising.

Other symptoms which should raise concern about neuroblastoma and prompt urgent referral include:

- proptosis
- unexplained back pain
- leg weakness
- unexplained urinary retention

In children or young people with symptoms that could be explained by neuroblastoma, an abdominal examination (and/or urgent abdominal ultrasound) should be undertaken, and a chest X-ray and full blood count considered. If any mass is identified, an urgent referral should be made.

Infants aged younger than 1 year may have localised abdominal or thoracic masses, and in infants younger than 6 months of age, there may also be rapidly progressive intra-abdominal disease. Some babies may present with skin nodules. If any such mass is identified, an immediate referral should be made.

2.2.5 Wilms’ tumour (all ages)

Wilms’ tumour most commonly presents with a painless abdominal mass. Persistent or progressive abdominal distension should prompt abdominal examination, and if a mass is found an immediate referral be made. If the child or young person is uncooperative and abdominal examination is not possible, referral for an urgent abdominal ultrasound should be considered. Haematuria in a child or young person, although a rarer presentation of a Wilms’ tumour, merits urgent referral.

2.2.6 Soft tissue sarcoma (all ages)

A soft tissue sarcoma should be suspected and an urgent referral should be made for a child or young person with an unexplained mass at almost any site that has one or more of the following features. The mass is:

- deep to the fascia
- non-tender
- progressively enlarging
- associated with a regional lymph node that is enlarging
- greater than 2 cm in diameter
A soft tissue mass in an unusual location may give rise to misleading local and persistent unexplained symptoms and signs, and the possibility of sarcoma should be considered.

These symptoms and signs include:

- **head and neck sarcomas:**
  - proptosis
  - persistent unexplained unilateral nasal obstruction with or without discharge and/or bleeding
  - aural polyps/discharge

- **genitourinary tract:**
  - urinary retention
  - scrotal swelling
  - bloodstained vaginal discharge

### 2.2.7 Bone sarcomas (osteosarcoma and Ewing’s sarcoma) (all ages)

Limbs are the most common site for bone tumours, especially around the knee in the case of osteosarcoma. Persistent localised bone pain and/or swelling requires an X-ray. If a bone tumour is suspected, an urgent referral should be made.

History of an injury should not be assumed to exclude the possibility of a bone sarcoma.

Rest pain, back pain and unexplained limp may all point to a bone tumour and require discussion with a paediatrician, referral or X-ray.

### 2.2.8 Retinoblastoma (mostly children aged under 2 years)

In a child with a white pupillary reflex (leukocoria) noted by the parents, identified in photographs or found on examination, an urgent referral should be made. The primary healthcare professional should pay careful attention to the report by a parent of noticing an odd appearance in their child’s eye.

A child with a new squint or change in visual acuity should be referred. If cancer is suspected, referral should be urgent, but otherwise referral should be non-urgent.

A family history of retinoblastoma should alert the primary healthcare professional to the possibility of retinoblastoma in a child who presents with visual problems. Offspring of a parent who has had retinoblastoma, or siblings of an affected child, should undergo screening soon after birth.
2.3 Investigations

When cancer is suspected in children and young people, imaging is often required. This may be best performed by a paediatrician, following urgent or immediate referral by the primary healthcare professional.

The presence of any of the following symptoms and signs requires investigation with full blood count:

- pallor
- fatigue
- irritability
- unexplained fever
- persistent or recurrent upper respiratory tract infections
- generalised lymphadenopathy
- persistent or unexplained bone pain (and X-ray)
- unexplained bruising.