Guidelines for Suspected Spinal Cord Compression in Children and Young People <18yr old
### Document Control

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**Information Reader Box**

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1 INTRODUCTION

Compression of the spinal cord and/or cauda equina is an oncological emergency that may lead to permanent, avoidable paralysis if not treated effectively.

Spinal cord compression can occur acutely in children with cancer. Rapid recognition of early signs and early, effective treatment maximise the chances of full recovery.

Cord compression can occur by a number of mechanisms including:
- Direct spread of tumour.
- Extension of tumour through vertebral foramina into epidural space
- Bony disease within vertebrae with secondary cord compression.

Sarcoma is the underlying diagnosis in the majority of cases, with neuroblastoma, lymphoma and leukaemia making up the bulk of the rest of cases. Occasionally leptomeningeal disease (e.g. metastatic medulloblastoma) can cause diffuse cord compression with less reliable localising signs. Primary spinal tumours and intraspinal metastases can also cause acute cord neurological dysfunction.

Cord compression may be the presenting feature of a new malignancy, or more frequently occurs as a consequence of metastatic disease progression or relapse.

Back pain occurs in up to 80% of children, and abnormal bladder or bowel function is common. Any patient with this combination of symptoms needs urgent investigation to rule out cord compression. Sensory abnormalities are less common, but need to be taken seriously when they occur. Patients (and their carers) who have a risk of spinal cord compression secondary to the site of the primary tumour should be made aware of such symptoms, and advised to seek urgent review if any develop.

A child with cancer who develops back pain should be considered to have spinal cord compromise until proved otherwise. The absence of weakness or sensory abnormalities does not reliably exclude the diagnosis.
2 MANAGEMENT OF SUSPECTED SPINAL CORD COMPRESSION

2.1 HISTORY

Take detailed history paying particular attention to the location and nature of pain, if present. Pain when straining to open bowels and pain at night are particularly concerning.

Ask about weakness, gait, bladder and bowel function (constipation, retention or incontinence) and any sensory abnormality (including perineal).

Establish the time since onset of symptoms and document the speed of progression.

2.2 EXAMINATION

Detailed neurological examination is mandatory.

Most patients with cord compression have weakness in distal muscle groups.

Sensory abnormalities and sensory levels are often more difficult to identify

The bony spine should be palpated for tenderness, which is a reliable localising sign if present.

Clinical examination often allows compressive lesions to be localised (See Table 1)

### TABLE 1 : CLINICAL LOCALISATION OF EPIDURAL CORD COMPRESSION

<table>
<thead>
<tr>
<th>SIGN</th>
<th>SPINAL CORD</th>
<th>CONUS</th>
<th>CAUDA EQUINA</th>
</tr>
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<tbody>
<tr>
<td>WEAKNESS</td>
<td>Symmetric, profound</td>
<td>Symmetric, variable</td>
<td>Asymmetric, May be mild</td>
</tr>
<tr>
<td>TENDON REFLEXES</td>
<td>Increased or absent</td>
<td>Increased knee,</td>
<td>Decreased, asymmetric</td>
</tr>
<tr>
<td></td>
<td></td>
<td>decreased ankle</td>
<td></td>
</tr>
<tr>
<td>BABINSKI</td>
<td>Extensor</td>
<td>Extensor</td>
<td>Plantar</td>
</tr>
<tr>
<td>SENSORY</td>
<td>Symmetrical</td>
<td>Symmetrical</td>
<td>Asymmetric Radicular</td>
</tr>
<tr>
<td></td>
<td>Sensory level</td>
<td>Saddle anaesthesia</td>
<td></td>
</tr>
<tr>
<td>SPHINCTER</td>
<td>Spared until late</td>
<td>Early involvement</td>
<td>May be spared</td>
</tr>
<tr>
<td>ABNORMALITY</td>
<td></td>
<td>Variable, may be</td>
<td>Variable, may be</td>
</tr>
<tr>
<td>PROGRESSION</td>
<td>Rapid</td>
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2. Management of suspected spinal cord compression
2.3 INVESTIGATION

Pre and Post Contrast MRI spine is the investigation of choice, and should be performed urgently, absolutely before 24 hours have elapsed. For children presenting in District General Hospitals with a strong suspicion of spinal cord compression, urgent transfer to a principal treatment centre is recommended to undertake further investigation and management. In some instances (for example, patients with advanced disease in the terminal phase of their illness) this approach may not be warranted and discussion of all cases with the Consultant Paediatric Haematologist/Oncologist is advised.

In most instances the whole spine is imaged. This is mandatory if there is any doubt about the level involved or there is a likelihood of multiple lesions (e.g. medulloblastoma). The conus should be included as this is a common site for metastatic central nervous system tumours, but may be difficult to image fully with MRI.

The case should be discussed with the Consultant Neuroradiologist on-call to arrange imaging. In addition, an anaesthetist will frequently be required in the case of young children.

Spine radiographs may be helpful to assess the vertebral bodies, although 50% of children will have normal x-rays.

Routine investigation should also include haematological and biochemical assessment (FBC, U&E, calcium, LFTs) to assess potential other disease processes and further complications.

2.4 MANAGEMENT

All patients must be discussed with Oncology/Haematology Consultant. The Neurosurgical team and Clinical Oncologists (Paediatric Radiotherapist) should also be included in these discussions.

Definitive Management
Management is complex and requires the input of a Consultant Haematologist/Oncologist in consultation with Neurosurgery and Clinical Oncology. The notes below explain some of the issues involved, but should not be seen as an alternative to a Consultant-led decision. Depending on the urgency and management route decided upon, it may be reasonable to wait until working hours before contacting the Clinical Oncology team.

Definitive management requires cord decompression which can be achieved in a number of ways; the most appropriate course of action depends on a number of factors including diagnosis (or lack of), speed of symptom progression and phase of illness e.g. presentation vs. terminal phase.

Dexamethasone can be given to reduce cord oedema associated with the compressive lesion. At best it can buy some time for imaging and allow definitive management to be arranged. The evidence underpinning dosing schedules is weak. If cord compression is suspected Dexamethasone (10mg/m2, max 16mg/day) may be given IV prior to emergency MRI scan. Dexamethasone should be continued onwards on the advice of the
neurosurgical/clinical or paediatric oncology teams, usually as a BD dose (although QDS may be considered) for two weeks.

If an epidural mass is demonstrated the spinal cord should be urgently decompressed if appropriate. Although corticosteroids reduce oedema rapidly and result in neurological improvement they are not an alternative to cord decompression.

Decompression may be achieved by surgery, local radiotherapy, or chemotherapy (depending on the underlying condition). In some children e.g. infants with neuroblastoma, or children with lymphoma, chemotherapy may be rapidly effective with fewer long-term side effects than other modalities.

When the diagnosis is uncertain, surgery provides an opportunity to biopsy the lesion in addition to treating the mass and may be the treatment of choice in good prognosis patients with surgically operable masses.

In children with a radiosensitive but not rapidly chemo-sensitive tumour, urgent radiotherapy is often preferable to other approaches. Dexamethasone is continued during radiotherapy.

**Please note:** management is complex and requires the input of a Consultant Haematologist/Oncologist. The notes above explain some of the issues involved, but are not an alternative to a Consultant-led decision.

**Supportive Management**

Children and young adults with suspected spinal cord compression require close and attentive supportive care. The following issues may need to be addressed:

- **Spinal stability.**
  Lesions infiltrating bone may significantly weaken the spine and until proven otherwise the child should be nursed flat with a neutral spine. Care of pressure areas is important

- **Pain.**
  Significant pain may require opiates or analgesia for neuropathic pain such as gabapentin.

- **Constipation.**
  Efforts should be made to maintain soft and regular bowel motions

- **Urinary retention.**
  Catheterisation may be required

- **Side effects of dexamethasone.**
  Hyperglycaemia, hypertension and gastric erosion may occur. Consider monitoring +/- H2 receptor blockers

- **Biochemical derangement.**
  Although uncommon, children and young people may suffer hypercalcaemia as part of an infiltrative process. They may also undergo tumour lysis and renal dysfunction secondary to dexamethasone therapy (in leukaemia/lymphoma).

- **Thromboprophylaxis.**
  Consideration should be given to use of stockings +/- LMWH in those at very high risk of thrombosis as per local guidelines.

Following treatment of spinal cord compression, discussion with physiotherapy should be undertaken to assess the need for, and manage, a rehabilitation programme.

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2. Management of suspected spinal cord compression
3 Further Information and References


NICE. Metastatic spinal cord compression. Diagnosis and management of adults at risk of and with metastatic spinal cord compression. CG 75 (2008)

