Early recognition and management of brain tumours in children


Abstract
Brain tumours comprise over one quarter of all childhood cancers in the UK and are the most common cause of cancer-related deaths in children. The presentation of brain tumours can vary substantially in children. The presenting symptoms are often similar to less serious conditions, and are often managed as such initially. Therefore, it can be difficult to diagnose brain tumours in children. An early diagnosis is usually associated with more effective treatment and improved health outcomes. The diagnostic interval between first presentation to a health professional and diagnosis for brain tumours in children has been shown to be three times longer in the UK than in other developed countries. As a result, the HeadSmart campaign launched a symptom card in 2011 to increase awareness of brain tumours in children among the general population and healthcare professionals, with the aim of reducing the diagnostic interval to 5 weeks. Nurses have an essential role in early recognition of brain tumours in children, and in providing care and support to the child and their family following a diagnosis.

Keywords
brain tumours, children, headache, HeadSmart campaign, neuroimaging, neuro-oncology

CANCER IS DIAGNOSED in 1,600 children under the age of 14 years in the UK annually (Children with Cancer UK 2016). Approximately 1 in 500 children under the age of 14 years are diagnosed with brain tumours each year in the UK (Wilne et al 2010), making them the second most common form of childhood malignancy after leukaemia (Children with Cancer UK 2016). Tumours of the central nervous system (CNS) (tumours of the brain and spinal cord) comprise approximately 26% of childhood malignancies, and brain tumours are the most common cause of cancer-related deaths in children in the UK (Children with Cancer UK 2016). Early recognition and management of brain tumours is essential to improve prognosis and minimise long-term complications for these children.

A brain tumour is an aberrant growth of brain cells that divide and replicate in an abnormal, uncontrolled way. The skull acts as a rigid container containing brain tissue, blood and cerebrospinal fluid in older children and adults (Paul et al 2013). The intracranial pressure increases as a result of increased tissue volume as the cells divide and the tumour grows. These changes may exert pressure on localised brain areas, compress cranial nerves or result in reduced perfusion to the brain, leading to ischaemia (Paul et al 2013). This article discusses early recognition of the signs and symptoms of brain tumours in children and the role of nurses in managing and supporting children and their families.

Diagnostic interval between presentation and diagnosis
A retrospective medical record review of 200 children with brain tumours over a 14-year period until June 2001 was undertaken in the Wessex Neurology Centre and Southampton General Hospital (Wilne et al 2006). This indicated that...
the diagnostic interval, the average time between first presentation and diagnosis, was approximately 2.5 months in the UK, representing a significant delay in the diagnosis of brain tumours in children (Wilne et al 2006). This was three times longer than the diagnostic interval in other developed countries, including North America, Poland, Israel and Switzerland (Wilne et al 2010). Symptoms may be experienced for varying lengths of time, ranging from 1 day to several years (Klitbo et al 2011). A delay in diagnosis is associated with suboptimal health outcomes in terms of morbidity, and long-term complications (Wilne et al 2010, Paul and Walker 2013, Royal College of Paediatrics and Child Health (RCPCH) et al 2011).

Recognition of the delay in diagnosis of brain tumours in the UK led to the launch of the ‘HeadSmart’ campaign in 2011. This aimed to reduce the time interval from presentation to diagnosis to 5 weeks, by increasing the awareness of brain tumours in children among the general population and healthcare professionals. A symptom card was introduced, entitled HeadSmart: Be Brain Tumour Aware (Figure 1) (RCPCH et al 2011), which describes specific symptoms for different age groups. This was designed to assist healthcare professionals in primary and secondary care by encouraging them to recognise relevant symptoms earlier and consider brain tumours as a possible diagnosis. The campaign led to a reduction in the total diagnostic interval to a median of 6.7 (mean: 21.3) weeks by May 2013 from a median of 14 (mean: 35.4) weeks in 2006 (HeadSmart: Be Brain Tumour Aware et al 2016). A free mobile version is also available for smartphones (www.headsmart.org.uk/headsmart-materials/).

Presentation of brain tumours in children
It can be difficult to identify brain tumours in children for two reasons. First, presentation in children may vary substantially, depending on the age of the child and the location of the brain tumour. Second, the signs and symptoms are often subtle and similar to less serious conditions (Table 1) (Wilne et al 2007, Wilne et al 2010, Paul and Walker 2013). It is essential that healthcare professionals use the HeadSmart symptom card.
card (Figure 1) (RCPCH et al 2011) as a guide and enquire about age-appropriate symptoms when a brain tumour is suspected (Paul et al 2014). Table 1 lists common presentations that healthcare professionals should be aware of and how they may be misdiagnosed in children with brain tumours.

**Clinical history and examination**

The signs and symptoms of a brain tumour are subtle. Therefore, it is essential to take a detailed, focused history of the patient and perform a thorough clinical examination. A health professional, ideally a paediatrician, should be involved if there is any suspicion that a child may have a brain tumour.

A retrospective cohort study involving 139 children with a brain tumour from four centres in the UK indicated that a median of one symptom or sign was reported at symptom onset, but that this increased to a median of six symptoms at the time of diagnosis (Wilne et al 2011). Therefore, healthcare professionals need to consider the possible diagnosis of a brain tumour at an earlier stage. They should monitor and investigate signs or symptoms that may be associated with a brain tumour in children, to enable the diagnosis of a brain tumour to be made before the tumour progresses. The child may then be referred to a specialist neurosurgical or oncology centre for further investigation.

A child’s neurological system should be assessed; however, it is important to note that a single neurological examination will not exclude a brain tumour (Wilne et al 2010). Tests that can be readily performed by healthcare professionals include: assessing the child’s gait by asking them to walk heel-to-toe in a straight line; assessing the child’s handwriting in school-aged children; measuring the child’s head circumference if they are under 2 years old; and assessing the child’s vision using a Snellen chart or other appropriate methods (Wilne et al 2006, Wilne et al 2010). Assessing vision can be difficult in young children, and it may be appropriate to refer the child to a paediatric ophthalmologist (Fry et al 2014).

Brain tumours may affect growth and the onset of puberty since they can alter the production and release of hormones from the pituitary gland. Therefore, healthcare professionals should record the child’s height and weight, and enquire about pubertal status in a sensitive and appropriate way (Wilne et al 2006, Wilne et al 2010).

Children should be referred for further investigations if their headache persists for over 4 weeks, and if other signs persist for over 2 weeks (Wilne et al 2013). A child with a history of migraine or tension headaches should be reviewed if the character of their headache changes, since these are known to mimic brain tumours. It can be difficult to assess headaches in children under the age of 4 years or in children with communication difficulties, because they may not be able to verbalise their headache. In these children, withdrawal and holding their head may indicate a headache (Wilne et al 2010).

**Clinical features**

Healthcare professionals should ask the patient and their parents about the symptoms listed in the HeadSmart symptom card (RCPCH et al 2011) and consider a review by a specialist medical professional.

### TABLE 1. Common presentations of children with brain tumours

<table>
<thead>
<tr>
<th>Presentation</th>
<th>Common misdiagnosis or misinterpretation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vomiting and nausea</td>
<td>Infective cause, viral gastritis.</td>
</tr>
<tr>
<td>Abnormal balance or gait</td>
<td>Otitis media, labyrinthitis.</td>
</tr>
<tr>
<td>Impaired growth and vomiting</td>
<td>Gastrointestinal disturbance.</td>
</tr>
<tr>
<td>Polydipsia and polyuria</td>
<td>Diabetes mellitus, rather than diabetes insipidus which may be associated with a brain tumour.</td>
</tr>
<tr>
<td>Changes in behaviour in young children</td>
<td>‘Badly behaved’, attention deficit disorder (ADHD), oppositional defiant disorder (ODD).</td>
</tr>
<tr>
<td>Headache and unilateral weakness</td>
<td>Hemiplegic migraine, which should be a diagnosis of exclusion, reached by a process of elimination.</td>
</tr>
<tr>
<td>Difficulty copying from the board at school</td>
<td>Refractive errors (often identified by an optician).</td>
</tr>
</tbody>
</table>

(Adapted from Wilne et al 2010, Fry et al 2014)
A child’s presentation is often age-dependent and it is vital that healthcare professionals consider the variation in manifestations when a child presents with a suspected brain tumour. Therefore, the HeadSmart symptom card (RCPCH et al 2011) (Figure 1) should be consulted when assessing a child with a suspected brain tumour. Table 2 summarises three UK studies on signs and symptoms of children with brain tumours at presentation and diagnosis. Nurses working in different environments will encounter children and young people presenting with a range of symptoms related to brain tumours and this information should be used in conjunction with the HeadSmart symptom card (RCPCH et al 2011). Children with headaches secondary to a brain tumour can have specific ‘red flag’ symptoms that

<table>
<thead>
<tr>
<th>Study</th>
<th>Number of participants</th>
<th>Age group</th>
<th>Symptoms at first presentation (%)</th>
<th>Symptoms at time of diagnosis</th>
<th>Clinical signs at diagnosis</th>
</tr>
</thead>
</table>
| Wilne et al (2006) | 200 patients presenting with a central nervous system (CNS) tumour. (The median symptom interval was 2.5 months.) | 15 weeks-17 years. | - Headache (41)  
- Vomiting (12)  
- Unsteadiness (11)  
- Visual difficulties (10)  
- Educational or behavioural problems (10)  
- Seizures (9) | | Neurological signs were present in 88% of cases at diagnosis:  
- Papilloedema (38)  
- Cranial nerve abnormalities (49)  
- Cerebellar signs (48)  
- Long tract signs (27)  
- Somatosensory abnormalities (11)  
- Reduced level of consciousness (12) |
| Wilne et al (2012) | 139 children with a brain tumour. (The median symptom interval was 3.3 months.) | 29 days-16.7 years. | - Headache (40, n=55)  
- Nausea and vomiting (28, n=39)  
- Motor system abnormalities (22, n=31)  
- Cranial nerve palsies (17, n=24)  
- Visual system abnormalities (23, n=17)  
- Endocrine or growth abnormalities (7, n=10)  
- Behavioural change (3, n=4) | - Headache (58, n=81)  
- Nausea and vomiting (63, n=88)  
- Motor system abnormalities (67, n=93)  
- Cranial nerve palsies (54, n=75)  
- Visual system abnormalities (69, n=96)  
- Endocrine or growth abnormalities (25, n=35)  
- Behavioural change (40, n=55) | |
| Chu et al (2015) | 181 patients with a primary intracranial tumour. | 0-24 years. | Clinical features in primary care were less specific. Visual disturbance in newborns to 4 year olds (rate: 0.49 per 100 person-months; 95% confidence interval (CI), 0.33-0.72). Headache in 5-24 year olds: 5-11 year olds (rate: 0.64; 95% CI, 0.47-0.88); 12-18 year olds (1.58; 95% CI, 1.21-2.08); 19-24 year olds (2.44; 95% CI, 1.71-3.49). | Predominant features at hospital admission were those of raised intracranial pressure in all age groups: From 1.17 per 100 person-months (95% CI, 1.08-1.26) in newborns to 4 year olds to 0.77 (95% CI, 0.67-0.88) in 19-24 year olds. |
suggest the presence of an intracranial mass which requires urgent investigation (Paul et al 2014). These red flag symptoms are listed in Box 1.

**Management**

**Emergency department management**

If a child presents to the emergency department with reduced consciousness or signs of raised intracranial pressure, the child should be triaged as ‘urgent’. Healthcare professionals need to act rapidly to stabilise the child using the ABCDE (airway, breathing, circulation, disability, exposure) approach. This should include assessment of their level of consciousness using the Glasgow Coma Scale (GCS), blood pressure and blood glucose (Advanced Life Support Group 2011). If the child's GCS score is below 15, high-flow oxygen should be administered, and if it is below 8, immediate airway protection is required (Paul et al 2014). Once the child is stabilised, their GCS should be reassessed every 15 minutes until their score normalises to 15. This reassessment should be continued when the child is relocated for further investigations, for example a computed tomography (CT) scan (Paul et al 2014).

**Radiological imaging**

Magnetic resonance imaging (MRI) is the most effective radiological imaging modality for diagnosing a brain tumour. However, a contrast-enhanced CT scan is often used in an emergency situation, or where a general anaesthetic for an MRI scan is not immediately available or is considered unsafe. If the investigation detects an intracranial lesion, the child should be referred urgently to neurosurgery specialists and will require further investigations as part of their diagnostic examination.

A management plan will then be developed, which may involve surgical resection, chemotherapy, and radiotherapy. Table 3 indicates preferred treatment regimens for common types of brain tumour (Lissauer and Clayden 2011). After a patient has completed treatment, they will require repeat MRI scans approximately every 6 months for 2 years, and then annually thereafter (Tidy 2015). Further detail on complex management regimens are beyond the scope of this article.

**Long-term management and prognosis**

Rehabilitation aims to improve overall physical, emotional, social, and educational outcomes, and has an important role during and after the child's treatment (National Institute for Health and Care Excellence (NICE) 2005). It may involve specialist nurses, neuropsychologists, physiotherapists, occupational therapists, and speech and language therapists (Wilne et al 2013).

The prognosis for children with brain tumours is variable, depending on where the tumour is localised, the type and stage of the tumour and the time taken to diagnosis. The overall 5-year survival rate is approximately 75% in children diagnosed with a brain tumour (Children with Cancer UK 2016). However, 60% of patients who survive have a life-altering disability (Wilne et al 2010). Possible long-term complications of CNS tumours include: epilepsy; ataxia; intellectual decline including reduced attention span and reaction time; speech difficulties; endocrine dysfunction, for example growth hormone deficiency; reduced bone density and an increased risk of subsequent tumours (Wilne et al 2010, Stocks et al 2012, Paul et al 2014).

Patients will require palliative care in cases where a cure is no longer possible. Therefore, children and their carers should have access to specialist paediatric

---

**BOX 1. ‘Red flag’ symptoms for headache associated with brain tumours**

- Abnormal head position, for example head tilt or stiff neck
- Unexplained headache in preschool children
- Change in level of consciousness
- New-onset severe or persistent headache, persisting for more than 4 weeks
- Occipital headache
- Vomiting, particularly in the morning
- Headache that awakens child in the night
- Headache that is worse at night or lying down
- Change in character of a previously diagnosed headache, for example migraine

(Paul et al 2014)
Challenges for children and their families

The diagnosis of a brain tumour has considerable effects on the child, their parents and family members. It is essential for healthcare professionals to communicate effectively with the child and the family, since care involves ‘a long and traumatic journey through many departments and hospital services’ (Sacree 2008). Where feasible, the multidisciplinary team should provide thorough preparation for the child and their family at diagnosis regarding surgery, rehabilitation and possible long-term disability or neurological deficit (Perrow 2013).

Research studies indicate that psychosocial support is beneficial in decreasing psychological distress throughout the course of the child’s illness (Steele et al 2015). Therefore, an effective information and support plan should be developed for the child and their family and should continue during the child’s treatment (Jones 2012). Ongoing support is essential, given the shock and uncertainty of diagnosis, the demands of care and the side effects of treatment. The cognitive and behavioural changes in the child that result from tumour growth can have a considerable effect on families, especially in children who are receiving palliative care (Zelcer et al 2010).

Families should be made aware of other sources of support that are available to them, for example support from CLIC Sargent and Macmillan Cancer Support. Families may also require financial support, since the costs of travel to specialist centres, care for other children and time off work can cause significant financial strain (Jones 2012, Fry et al 2014). Healthcare professionals should ensure that families are aware of the Disability Living Allowance benefit and potential bursaries and charities that may be able to offer financial support.

Role of the nurse

Nurses have a central role in diagnosing, managing, and supporting children with brain tumours and their families.

School nurses and nursery nurses

School nurses and nursery nurses often see the same children over a prolonged period and are well-placed to identify possible signs of a brain tumour. Changes observed by nursery nurses or school nurses may include a delay in growth, tiredness, frequent absences from school, behavioural changes, a request for analgesics for frequent headaches and frequent falls (Wilne et al 2010). They should alert the child’s parents if they are concerned.

Practice nurses

Nurses working in primary care frequently care for children in their clinical practice and may identify signs or symptoms of brain tumours in children that are coincidental to their presenting symptoms. Young children may also be seen by nurses in primary care because they are experiencing visual difficulties. Therefore, it is essential that these nurses are aware of the signs and symptoms of brain tumours in children of different ages, as indicated on the HeadSmart symptom card (RCPCH et al 2011). If there are any symptoms that cause concern, the practice nurse should take a detailed history, examine the child if appropriate and discuss the patient with a GP in the practice who may decide to monitor the child’s symptoms or refer them for further investigations.

<table>
<thead>
<tr>
<th>TABLE 3. Preferred management options for brain tumours</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Type of brain tumour</strong></td>
</tr>
<tr>
<td>Astrocytoma</td>
</tr>
<tr>
<td>Craniopharyngioma</td>
</tr>
<tr>
<td>Medulloblastoma</td>
</tr>
<tr>
<td>Brainstem glioma</td>
</tr>
</tbody>
</table>

(Lissauer and Clayden 2011)
Emergency department nurses

Nurses in the emergency department have a crucial role in urgently assessing children who present with reduced consciousness and ensuring they are stabilised (Paul et al 2014). These nurses should be aware of the appropriate management of a child who presents with reduced consciousness or signs of raised intracranial pressure (Box 2) and their role in this.

Some children presenting to the emergency department may have a subtle presentation, for example children who have simple injuries resulting from recurrent falls that are secondary to new onset gait abnormalities. In some instances, the child may have experienced a loss of balance or a change in their vision as a result of a brain tumour. Therefore, nurses should always consider an intracranial lesion as a differential diagnosis, and should enquire about risk factors and the signs and symptoms on the HeadSmart symptom card during triage (Paul et al 2014).

Children who are receiving chemotherapy treatment following their diagnosis of a brain tumour may present to the emergency department with febrile neutropaenia. Febrile neutropaenia is a well-recognised cause of death in children with cancer and should be considered an emergency (Bate et al 2013). Nurses should be aware of this complication, which results from sepsis. Children with febrile neutropaenia may present with hypotension, tachypnoea, hypoxia (oxygen saturations less than 94%), severe mucositis, vomiting, abdominal pain, focal infection and a reduced neutrophil count (<0.1x10⁹/L) (Bate et al 2013). Nurses should ensure that there is no delay in administering intravenous antibiotics to these children (Bate et al 2013).

Children and young people’s nurses

Children diagnosed with brain tumours have frequent contact with children and young people’s nurses throughout their treatment and thereafter. Children and young people’s nurses are the main group of healthcare professionals responsible for providing ongoing support and day-to-day care for these children while they are in hospital. Their role includes: coordinating the child’s investigations and management; ensuring the child is monitored correctly on the ward; recording paediatric early warning scores (PEWS); administering medications and chemotherapy; and reviewing the child regularly to identify any new signs or symptoms or changes to existing symptoms.

Children and young people’s nurses may spend considerable time with individual patients and their families, and they often develop a close relationship with them. These nurses have an important role in providing support to the child and family, and they are also well-placed to learn the family dynamics. Nurses can also link the child and family to available support networks. Depending on family circumstances, further support could include: referring them to support services, arranging on-site accommodation for families travelling to specialist centres, arranging care for siblings through social services or guiding the families to sources of financial support. Children and young people’s nurses are well placed to identify the support needed for families, which will be unique to that particular family and should contribute to multidisciplinary team meetings to ensure delivery of an appropriate individualised care package (Perrow 2013).

**Conclusion**

Brain tumours are the second most common form of childhood malignancy after leukaemia and are the most common cause of cancer-related deaths in children in the UK. Healthcare professionals should be aware of the possible signs and symptoms of a brain tumour in children of different ages, and should consider a brain tumour...
as a potential differential diagnosis for common presentations that may appear to have a slightly different presentation from that which is expected for such illnesses. Nurses in many practice settings are in a position to recognise possible symptoms and signs of brain tumours in children and to raise concerns earlier. This is likely to reduce the time from presentation to diagnosis, which will improve the prognosis for these children and reduce the likelihood of long-term complications.

References


