20.1 Spinal Cord Compression

20.1.1 Incidence
Spinal cord compression (SCC) is uncommon in children, occurring in 2.7–5% of children with cancer and 4% of children at diagnosis of cancer (Kelly and Lange, 1997).

SCC is most common in terminal stages of metastatic cancer, but 25–35% of cases occur as a presenting complaint, usually due to extension of a paravertebral neuroblastoma, Ewing’s sarcoma, non-Hodgkin’s lymphoma (NHL), or Hodgkin’s disease through one or more intravertebral foramina – known as the “dumb-bell tumor” (Nicolin, 2002). SCC can also present as a manifestation of tumour recurrence that is most commonly seen in children with rhabdomyosarcoma or osteosarcoma.

20.1.2 Etiology
SCC occurs when extension of a primary tumour causes compression of the vertebral venous plexus, leading to

- Vasogenic oedema
- Venous haemorrhage
- Demyelination
- Ischaemic cell death

Presenting features of SCC in children include

1. Back pain
   - Localised or radicular
2. Weakness
   - Ambulatory
   - Nonambulatory
   - Paraplegic
3. Localised spine tenderness
4. Sphincter disturbances – usually urinary retention or constipation
5. Sensory disturbances (difficult to ascertain in children)
6. Gait disturbances

(Nicolin, 2002; Kelly and Lange, 1997)

Back pain is unusual in children and should be investigated promptly. Pain may be aggravated by movement, neck flexion, or a recumbent position.

Magnetic resonance imaging (MRI) is the current initial investigation required because radiographs are abnormal in only one-third of cases (Parisi et al., 1999; Nicolin, 2002). MRI gives high-quality images of the spinal cord, epidural space, and paravertebral areas.

If possible, lumbar puncture (LP) should not be performed when SCC is suspected, due to the risk of impaction of the cord (spinal coning). If LP is necessary, close neurological monitoring is essential.

20.1.3 Treatment

The goal of emergency treatment is to restore neurological function and avoid irreversible damage. Intravenous dexamethasone 1 mg/kg infused over 30 minutes should be administered. Cases without neurological deficits may be given a lower dose of dexamethasone, 0.25–0.5 mg/kg orally every 6 hours. Doses are empiric, and large doses of dexamethasone are not justified (Kelly and Lange, 1997).

Surgical debulking of the tumour may be necessary through laminectomy (removal of the posterior arch of the spinal canal) (Acquaviva et al., 2003). These procedures often lead to later problems, with further reconstructive spinal treatments required. Osteoplastic laminotomies, followed by bracing for 6–8 weeks, are now the preference of some surgeons. Long-term incidence of deformities with this procedure has not yet been established.

If there is a known diagnosis, radiotherapy can be used in radiosensitive tumours (Nguyen et al., 2000). Low-dose radiotherapy is recommended because spinal radiation >2,000 cGy can cause late scoliosis and kyphosis, particularly in young children.

Chemotherapy is also effective in relieving pressure on the spinal cord from chemo-sensitive tumours, such as neuroblastoma, NHL, Hodgkin’s disease, and Ewing’s sarcoma. Chemotherapy has the advantages of avoiding long-term deformities and gaining control of the cancer at the primary or metastatic sites.

20.1.4 Prognosis

The ultimate outcome for these children depends on

- The extent of cancer at diagnosis
- The response to treatment

Quality of life, however, depends on neurological recovery, which is related to the degree of disability at diagnosis. In turn, this is associated with the duration of symptoms and time to diagnosis. Patients who are ambulatory at diagnosis generally remain ambulatory, and about half of the children who are nonambulatory at diagnosis regain ability (Kelly and Lange, 1997). Immediate treatment is essential.

20.2 Fatigue

A study carried out at two major cancer centres in the southern United States arrived at a definition for fatigue from focus group sessions: Fatigue is a profound sense of being tired or having difficulty with movement, such as using arms and legs or opening eyes, and is influenced by environmental, personal/social, and treatment-related factors and can result in difficulties with play, concentration, and negative emotions, most typically anger and sadness (Hockenberry-Eaton et al., 1999). The profound sense of tiredness can be acute, episodic, or chronic, and is relieved by rest and distraction.

20.2.1 Incidence

Fatigue during treatment for cancer in childhood and adolescence is now accepted as being a near-universal experience that adversely affects the quality of life of patients and their families. Fatigue is reported by clinicians to be particularly common in children and adolescents with leukaemia. Healthy people seldom
regard fatigue as a serious problem because it is usually a temporary phenomenon; however, for those with cancer, it is a chronic and frequently relentless symptom.

The actual incidence of fatigue in children or adolescents treated for cancer is unreported, although Bottomley et al. (1995) found that over 50% of school-age children (n=75) with cancer reported being tired and playing less than they did before the illness. It is acknowledged that fatigue is probably underrecognised and undertreated despite its being a prevalent problem in the paediatric population (White, 2001).

### 20.2.2 Etiology

The etiology of this type of fatigue is complex. There can be many contributing factors in the cause of this type of fatigue:

1. **Physiological**
   - Physiological causes include anaemia, nutritional status, and biochemical changes secondary to disease and treatment. Fatigue may be attributed to bone marrow transplantation, surgery, radiation, or/and chemotherapy. Treatment for childhood cancer is aggressive, with every effort made to administer maximum doses of therapy when possible. Unlike adult regimens, dose-limiting parameters do not include fatigue as a side effect.
   - Young children may be unaware of changes in their physical stamina and activities of daily living, while parents and older children/adolescents may simply accept their fatigue and lack of energy as a consequence of having cancer.

2. **Psychological**
   - Anxiety and depression can lead to fatigue (Langeveld et al., 2000). These are complex issues because fatigue may be due to a depressed mood, or people may become depressed if they perceive that they are constantly fatigued (Langeveld et al., 2000). Additionally, depression and fatigue may co-occur in cancer patients, as they can both originate from the same pathology (Visser and Smets, 1998).

3. **Situational**
   - Sleep patterns are very frequently changed, especially during stays in hospital, which again can be a contributing factor to feelings of general lethargy and tiredness.

The causes of fatigue and their contributing factors identified by children are listed in Table 20.1. Adolescents recognised the following as reasons for fatigue:

- Noise
- Inability to sleep
- Feeling upset
- Fear
- Effects of treatment
- Boredom

(Hockenberry-Eaton et al., 1999)

Parents, however, stated the following factors:

- Hospital sounds
- Interruptions
- Waiting
- Needing to interact with too many other individuals

<table>
<thead>
<tr>
<th>Cause of fatigue</th>
<th>Contributing factors</th>
</tr>
</thead>
<tbody>
<tr>
<td>Treatment</td>
<td>Chemotherapy, radiation, surgery</td>
</tr>
<tr>
<td></td>
<td>Fatigue associated with being sick</td>
</tr>
<tr>
<td>Being active</td>
<td>Easily tired after play and activities</td>
</tr>
<tr>
<td>Pain</td>
<td>Being tired when experiencing discomfort such as pain</td>
</tr>
<tr>
<td>Hospital environment</td>
<td>Noises, frequent interruptions, location of bed. Trouble falling asleep</td>
</tr>
<tr>
<td>Sleep changes</td>
<td>Sleep patterns change making it hard to get to sleep or sleep all night</td>
</tr>
<tr>
<td>Low counts</td>
<td>Feeling tired when experiencing myelosuppression</td>
</tr>
</tbody>
</table>
A study by Davies et al. (2002) found that children with cancer may experience three subjectively distinct types of tiredness: typical tiredness, treatment fatigue, and shutdown fatigue.

### 20.2.3 Treatment

Because fatigue is commonly unrecognised in this patient population, interventions need to begin with an educational component that provides patients, parents, and staff with critical information about diagnosing fatigue and describing the type of fatigue experienced. It is only once the type of fatigue is identified that interventions can then be suggested. Precursors to fatigue identified in various studies have included physical, environmental, mental, and psychological causes that have implications for clinical care (Davies et al., 2002).

Cancer and cancer treatment can place an increasing and extraordinary demand on the child’s energy. Fatigue may also be an issue related to mental health for paediatric oncology patients. Some of the common symptoms of fatigue may be misinterpreted as indications of depression. After careful nursing assessment for signs of fatigue versus depression, the child/adolescent may need to be referred to the mental health team. The relationship between nutrition and fatigue is also a concern because inadequate nutrition and anorexia can affect the child’s energy levels. Efforts to optimise nutritional status can help support children through the potential for fatigue.

An improved understanding of the contributing and alleviating factors associated with fatigue in this patient population will provide children with greater comfort during treatment. Within the plethora of information the families receive during treatment of their child’s cancer, it is important for the nurse to discuss fatigue as a symptom both during and after treatment. Awareness of interventions that will decrease fatigue can also be discussed, both for the hospital and for the home. In the study by Hockenberry-Eaton et al. (1998), children and adolescents reported factors that may help overcome fatigue (see Table 20.2).

The realisation that the hospital environment can be a major contributor to fatigue in children and adolescents is important. Awareness that fatigue during hospitalisation occurs because of disrupted sleep due to noises, frequent interruptions, and even the location of the room can stimulate thoughts and ideas on how to make the hospital setting more conducive to rest and sleep.

Table 20.3 gives examples of nursing interventions that can relate to fatigue-alleviating factors as described by Hinds and Hockenberry-Eaton (2001).

### 20.2.4 Prognosis

Any effort to define, measure, and intervene with fatigue needs to take into consideration the major components of these children and adolescents’ treatment context. Fatigue is a problem for many long-term survivors of childhood cancer and may be multifactorial in nature. Fatigue in these patients may be associated with certain late effects of chemotherapy, including irreversible cardiac and pulmonary toxicities. Efforts should focus on educating survivors to avoid factors that may contribute to fatigue, including those behaviours that may potentiate organ toxicity, such as tobacco and alcohol use (Hollen and Hobbie, 1993).

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**Table 20.2.** Children and adolescents description of what helps overcome fatigue (Hockenberry-Eaton et al., 1999)

<table>
<thead>
<tr>
<th>Reported methods of help to overcome fatigue</th>
<th>Explanation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Naps/sleep</td>
<td>Resting during the day and night</td>
</tr>
<tr>
<td>Visitors</td>
<td>Someone coming to visit may help</td>
</tr>
<tr>
<td>Fun/activities</td>
<td>Going to the movies/listening to music/reading a book</td>
</tr>
<tr>
<td>Blood transfusion</td>
<td>Can give back some energy</td>
</tr>
<tr>
<td>Protected rest time</td>
<td>Not getting interrupted during rest times</td>
</tr>
<tr>
<td>Going outside</td>
<td>Outside to enjoy the day and get some fresh air</td>
</tr>
<tr>
<td>Having fun</td>
<td>Doing something they like in hospital and at home</td>
</tr>
</tbody>
</table>
Most young children seem to recover their energy levels fairly quickly even between pulses of chemotherapy, whereas adolescents seem to take much longer. Those who seem at risk of trying to fight off fatigue in the long term are those adolescents who have had some form of megatherapy. Follow-up treatment for these patients can be lengthy, requiring many outpatient visits, which in turn can lead to psychological and physical distress that may manifest itself as increasing fatigue.

As future work is carried out in the whole area of fatigue in paediatric oncology patients, we should improve our understanding of the individuals’ experiences and ultimately provide them with a greater sense of understanding and comfort during treatment for their cancer.

### 20.3 Cognitive Deficits

#### 20.3.1 Incidence

It is now generally accepted that central nervous system (CNS) treatments for childhood cancer can result in significant cognitive impairment, most commonly in the areas of attention/concentration.

Kingma et al. (2000) found that magnetic resonance imaging (MRI) of the brain revealed abnormalities in 63% of cases of children treated for acute lymphoblastic leukaemia (ALL) who received cranial irradiation and intrathecal methotrexate. The resultant cognitive impairment is commonly manifested as lower intelligence (IQ) and memory capacity and poorer academic achievement and visual-motor functioning. It remains unclear whether these deficits stabilise or diminish with time since treatment or if there may be an ongoing decline in abilities.

Altered mental status is frequently observed in children being actively treated for systemic cancer. However, the majority of these patients suffer from iatrogenically-induced encephalopathy, predominantly opioid-related.

Children treated at or before 5 years of age are considered to be at high risk with respect to neurobehavioral impairment, as researchers believe that the developing central nervous system may be particularly vulnerable to toxic agents during this time. The nature of the deficits suggests that children treated with cranial radiotherapy and chemotherapy are able to learn but do so more slowly than other children.

### Table 20.3. Nursing interventions in the alleviation of fatigue (Hinds and Hockenberry-Eaton, 2001)

<table>
<thead>
<tr>
<th>Factor</th>
<th>Interventions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hospital environment</td>
<td>Decrease unit noise levels</td>
</tr>
<tr>
<td></td>
<td>Group nursing activities together</td>
</tr>
<tr>
<td></td>
<td>Implement protected rest times</td>
</tr>
<tr>
<td></td>
<td>Maintain quiet hours at the nurses’ station</td>
</tr>
<tr>
<td>Personal/behavioural</td>
<td>Establish a routine/schedule in the hospital setting</td>
</tr>
<tr>
<td></td>
<td>Offer choices in relation to care where possible</td>
</tr>
<tr>
<td></td>
<td>Provide activities to prevent boredom in the hospital setting</td>
</tr>
<tr>
<td></td>
<td>Encourage participation of care in a positive manner</td>
</tr>
<tr>
<td>Treatment-related</td>
<td>Assess the need for blood transfusion</td>
</tr>
<tr>
<td></td>
<td>Consider physical exercise as part of the daily hospital schedule</td>
</tr>
<tr>
<td></td>
<td>Support nutritional needs</td>
</tr>
<tr>
<td></td>
<td>Manage other side effects that may enhance fatigue</td>
</tr>
<tr>
<td>Cultural/family/other</td>
<td>Educate families on the symptom of fatigue</td>
</tr>
<tr>
<td></td>
<td>Inform parents that children/adolescents receive cues from their behaviour</td>
</tr>
<tr>
<td></td>
<td>Promote visits by family and friends</td>
</tr>
<tr>
<td></td>
<td>Encourage quiet activities that expend minimal energy</td>
</tr>
</tbody>
</table>

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Most young children seem to recover their energy levels fairly quickly even between pulses of chemotherapy, whereas adolescents seem to take much longer. Those who seem at risk of trying to fight off fatigue in the long term are those adolescents who have had some form of megatherapy. Follow-up treatment for these patients can be lengthy, requiring many outpatient visits, which in turn can lead to psychological and physical distress that may manifest itself as increasing fatigue.

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### 20.3 Cognitive Deficits

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20.3.2 Etiology

There is well-documented evidence that children who have received cranial irradiation for treatment of ALL are likely to have resulting cognitive deficits (Moore et al., 2000; Precourt et al., 2002).

Neuroradiological signs in ALL survivors include brain abnormalities such as calcification, white matter changes, and parenchymal atrophy. The underlying cerebral pathology includes necrotising leucoencephalopathy and mineralising microangiopathy.

Organicity in behavioural sequelae for survivors, documented as lack of initiative, loss of motivation, increased distractibility, flat affect, and irritability, has been described as similar to that of patients with frontal lobe abnormalities.

The most important antecedent factors precipitating cognitive late effects in long-term survivors of childhood cancer are listed in Table 20.4.

20.3.3 Treatment

Cranial irradiation has been eliminated from most treatment protocols. However, patients with meningeal leukaemia or those receiving bone marrow transplantation may be confronted with academic limitations. Initial studies have also shown that dexamethasone therapy (compared with prednisolone) may increase the risk for neurocognitive effects in children treated for ALL (Waber et al., 2000).

A study done in the United States found that cancer-surviving adolescents may require intervention to improve their decision-making skills (Hollen et al., 1997). Poor-quality decision making was also clearly linked to adolescents who exhibited more risk behaviours.

Particular attention should therefore be paid to the development of concentration, attention, short-term memory, and abstract reasoning ability in all children, with the development of verbal processing skills needing greater attention in girls. Deficits in abstract reasoning, problem solving, and planning ability have also been found in survivors of childhood cancer.

Time missed from school is likely to be correlated with the degree of illness and medical complications, making periods of absence unavoidable. But when children are well, parents should be encouraged to send them to school as often as possible.

20.3.4 Prognosis

As the number of children surviving cancer for extended periods of time continues to increase, the phenomenon of symptoms that persist following completion of treatment is being recognised. Some children may benefit from special educational assistance to improve their educational outcomes. This is important in order to improve the quality of life of survivors and to help them achieve their maximum potential, initially at school and ultimately in the workforce.
20.4 Diabetes Insipidus

20.4.1 Incidence

Diabetes insipidus (DI) is found postoperatively in patients with craniopharyngioma. It is also the most common clinical presentation of patients with CNS disease in Langerhans cell histiocytosis (LCH). The clinical features of polyuria and polydipsia are usually dramatic. However, formal confirmation of the diagnosis by a water deprivation test is recommended because of occasional confusion with psychogenic polydipsia. DI may also be a late sequela of LCH, as found in one study in which 25% of patients followed up more than 3 years after diagnosis were on treatment (Broadbent and Pritchard, 1997). DI may also be one of the presenting features in intracranial germ cell tumours (Tarng and Huang, 1995).

20.4.2 Etiology

DI is characterised by polyuria and polydipsia, due to a disorder of antidiuretic hormone (ADH) availability. There are two types:-

1. Central or neurogenic DI: a defect in the synthesis or release of ADH
2. Nephrogenic DI: failure of the kidneys to respond to ADH

An abnormal growth hormone response has been correlated with the presence of DI in some studies. The diagnosis of DI is based on the results of a water deprivation test with measurement of urinary arginine vasopressin (AVP). The DI may be termed as “complete” or “partial.”

20.4.3 Treatment

The aim of treatment is to treat any underlying disorder and supply the body with pharmacologic preparations that contain the missing hormone. These preparations cannot be given orally because the gastrointestinal tract destroys them; they must be administered parenterally or nasally. The preferred drug for treating chronic DI is 1-desamino-8-D arginine vasopressin (DDAVP). This can be given by intranasal spray and has a duration of action of 8–20 hours.

20.4.4 Prognosis

The most appropriate treatment for reversing DI-complicating LCH is yet to be determined (Broadbent and Pritchard, 1997). The reports of one study that looked at the endocrine sequelae of childhood craniopharyngioma found that most children developed DI postoperatively (Bin-Abbas et al., 2001). Patients with an initial diagnosis of idiopathic DI require vigilant medical follow-up including repeated neuroimaging studies, particularly when there is evidence of evolving pituitary hormone deficiencies.

References


