Neck lumps and head and neck tumours in children

This article discusses the presentation, investigation and treatment of both benign and malignant lumps encountered in the head and neck region in children.

Neck lumps in children are common and are mostly secondary to reactive lymphadenopathy. These are usually self-limiting but may progress to abscess formation requiring incision and drainage. Chronic infections are less common and congenital abnormalities can present as a neck swelling at birth or frequently at a later date following an acute infective episode. Head and neck malignancies in children are rare. Many present with an asymptomatic swelling in the head and neck region. Survival rates have significantly improved over the past 20 years and it is therefore particularly important to detect these at an early stage. This article will review the presenting symptoms and signs of a variety of pathologies in the head and neck region in children and will indicate the appropriate investigations and treatment regimens.

Head and neck anatomy

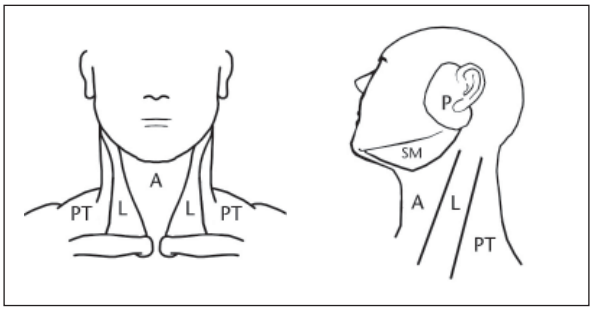
In the neck, the sternomastoid muscle on each side divides the area into anterior and posterior triangles (*Figure 1*). The submandibular triangle lies superior to the digastric muscle and below the mandible. Lymph nodes are distributed throughout the neck but the main deep cervical chain runs down almost parallel to and deep to the sternomastoid.

The site of any swelling can indicate the likely underlying diagnosis (*Table 1*).

General principles of history taking

Establish whether the swelling is acute (less than 6 weeks) or has become chronic (greater than 6 weeks). Swellings that are apparent at, or shortly after birth are likely to be congenital although some congenital anomalies may not become apparent until later. Symptoms suggestive of upper respiratory tract infection should be sought (fever, rhinorrhoea, malaise). Contact with persons with known infection (e.g. tuberculosis) should be established as should a history of overseas travel or exposure to farm animals, cats and ticks.

Figure 1. Triangles of the neck. A = anterior; L = lateral (sternomastoid); P = parotid; PT = posterior; SM = submandibular.



General principles of examination

The swelling itself should be carefully assessed with particular attention to the location in the neck, its size, its

Table 1. The distribution of masses in the neck

Location of swelling in neck	Pathologies encountered in order of frequency
Lateral neck	Enlarged lymph nodes
	Branchial cysts
	Lymphangiomas and haemangiomas
Central	Thyroglossal duct cysts
	Dermoid cysts
	Enlarged lymph nodes
	Thyroid masses
Parotid region	Mumps
	Bacterial parotitis (often recurrent in children)
	Vascular or lymphatic malformations
	Branchial anomalies
	Enlarged intra-/extra-parotid lymph nodes
	Parotid neoplasms
Submandibular region	Enlarged lymph nodes
	Floor of mouth infection or dental abscess
	Lymphatic or vascular malformation
Posterior triangle	Enlarged lymph nodes
	Branchial anomalies
	Lymphatic malformations

Malignant lesions are rare but can occur in any of these locations

Miss FB MacGregor *is Consultant Otolaryngologist, Royal Hospital for Sick Children, Yorkhill, Glasgow G3 8SQ and* **Miss KA McAllister** *is FTSTA 3 Otolaryngology, Gartnavel General Hospital, Glasgow*

Correspondence to: Miss FB MacGregor

mobility and whether it is red, tender and hot. Fluctuation indicates a cystic mass or abscess formation. A careful examination of the remainder of the head and neck region should be made with particular regard to possible sources of infection or sites of a primary malignancy in suspicious cases. These include the oropharynx, oral cavity, teeth, nose and ears. Examine the scalp and skin in addition.

Cervical lymphadenopathy

Children are predisposed to frequent upper respiratory tract infections and subsequent enlargement of cervical lymph nodes is very common. The huge majority of enlarged lymph nodes are reactive and serious pathology is rare. However, it is important to exclude an underlying malignancy in children who present with cervical lymphadenopathy and there are certain features which are associated with a high risk of serious pathologies. These are lymph nodes in the supraclavicular area, lymph nodes which measure more than 3 cm, lymph nodes in children with a history of malignancy and children who have other findings such as hepato-splenomegaly, fever, weight loss or night sweats (Kubba, 2006). In these cases urgent excisional biopsy is recommended.

When the diagnosis is less clear then a number of investigations may be implemented. A chest X-ray may be appropriate and bloods should be taken for full blood count, infectious mononucleosis, Bartonella (cat scratch), toxoplasmosis and cytomegalovirus in persistent cases. A Mantoux test may be indicated. An ultrasound scan can be useful in determining whether the lymph node has a normal architecture or it may raise the suspicion of malignancy. Fine needle aspirate cytology is generally unhelpful in a paediatric setting and when there is reasonable concern, excision biopsy is recommended (Hartley, 2007).

Acute infective lymphadenopathy is usually self-limiting or settles with antibiotic therapy but abscess formation may occur and require incision and drainage.

Atypical mycobacterial infection

Atypical mycobacterial infection resulting in chronic cervical lymphadenopathy is not uncommon. Initially children will present with painless enlarged neck nodes (lymphoma may be suspected) but with time the overlying skin becomes red and indurated. Abscess formation can occur resulting in a persistent discharging sinus (*Figure 2*). Diagnosis may be made by fine needle aspirate cytology or excision biopsy. A chest X-ray and Mantoux should be performed (Hartley, 2007).

Ideally complete surgical excision is performed to eradicate the disease but where access is limited or surgery is contraindicated because of significant risk of damage to surrounding structures (e.g. the facial nerve) then the use of antituberculous therapy should be considered.

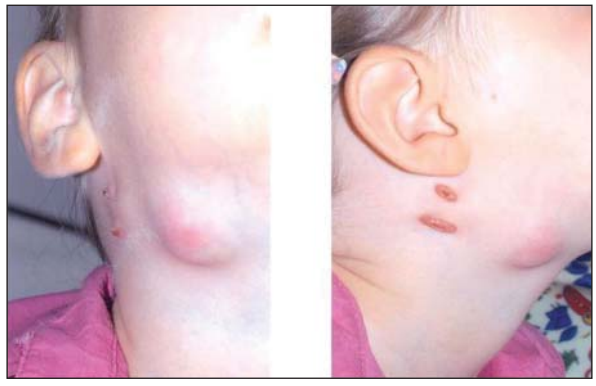


Figure 2. Atypical mycobacterial infection – submandibular swelling, parotid swelling and two discharging sinuses at previous incision and drainage sites.

Congenital cysts and other developmental anomalies

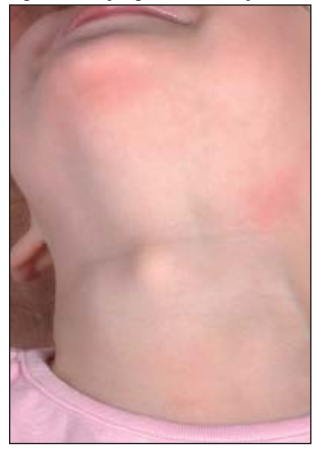
Developmental anatomy of the head and neck region is complex. Some understanding of this is vital to be able to recognize a number of congenital anomalies that present in childhood (MacGregor, 2007). Failure of this process to proceed normally predisposes children to cysts, sinus and fistula formation in the neck.

Thyroglossal duct cyst

At approximately 7 weeks in utero the thyroid gland descends from the foramen caecum at the base of the tongue to sit in front of the primitive foregut. The duct that descends behind this should involute with time but if this does not occur then a cyst may develop anywhere along this anatomical pathway. Children or young adults can present with a midline smooth fluctuant swelling in the neck usually closely associated to the hyoid bone (*Figure 3*). This can become infected and present with abscess or fistula formation (Maddalozzo et al, 2001). Typically, any cyst will move on swallowing or protrusion of the tongue.

Any infection should be treated and the investigation of choice is an ultrasound scan to confirm that there is a normal thyroid gland separate to this (to avoid the rare possibility of the excision of the only functioning thyroid tissue in the neck). Histological sections have confirmed that the duct or tract may be very closely associated with the hyoid bone. Therefore excision should include not just the cyst but the middle portion of the hyoid bone and a wedge of muscle extending up to the foramen caecum. This is called a Sistrunk's procedure.

Figure 3. Thyroglossal duct cyst.



Dermoid cyst

Dermoid cysts in the neck are often mistaken for thyroglossal duct cysts. They are formed by fusion of midline epithelial elements in utero. They are smooth discrete mobile cysts filled with sebaceous material and are often found in the midline of the anterior neck. Simple excision is the treatment of choice.

Nasal dermoid cyst

Nasal dermoids also occur in the midline anywhere from below the tip of the nose up to the forehead. These are usually associated with a punctum often over the bridge of the nose and hairs may extrude from this. These frequently become infected and require excision. The challenge in this situation is that the tract or indeed a cyst can occur anywhere up to the skull base around the area of the crista galli. Imaging is therefore important before embarking on surgery (computed tomography (CT) scanning and MRI scanning). Surgical excision is the treatment of choice (Bloom et al, 2002).

Branchial apparatus anomalies

In early utero a series of arches with external clefts and internal pouches become apparent and failure to develop normally can result in a cyst, fistula or sinus occurring in the head and neck region (Salder, 1985). Often these do not become apparent until infection occurs. Because these lesions are rare there is often a delay in diagnosis and some children have undergone multiple surgical interventions before the underlying problem is recognized.

First branchial arch anomaly

These often present with a cyst or mass in the periauricular area or upper neck just anterior to the sternomastoid. A tract leading to the external auditory canal may be present and a child can present with a discharging sinus in the external auditory canal. Diagnosis is often delayed (Ford et al, 1992). Investigations will often include a CT scan or MRI scan and the treatment is excision after infection has settled. These lesions can be closely associated with the facial nerve and surgery should be carried out by an experienced surgeon with the aid of a facial nerve monitor.

Third and fourth branchial pouch anomalies

These are rare and are often diagnosed late (Rea et al, 2004). Neonates can present with huge cysts requiring emergency airway intervention. Alternatively, older children can present with a recurrent thyroiditis or anterior neck abscess (*Figure 4*) as they have a sinus extending down from the pyriform sinus to the upper pole of the thyroid gland. Management includes treatment of the infection and then a barium swallow to confirm the presence of a sinus. The treatment of choice is excision of the tract although direct cautery to the sinus opening is now being advocated in some departments.

Branchial cysts

It is not entirely clear whether these lesions which occur as a cystic swelling anterior to sternomastoid in the upper neck are the result of failure of the cervical sinus to close in utero or caused by a cyst forming within the lymph nodes (*Figure 5*). However, the management involves treatment of any infection and complete excision of the cyst.

Lymphatic malformations

Lymphatic malformations or cystic hygromas can present at birth or develop during childhood (Hartley, 2007). The swellings consist of dilated lymphatic channels and associated epithelial-lined cysts. The cyst may be large (macrocystic) or small (microcystic) or a combination of the two. They are commonly associated with venous malformations. These can occur in the neck, floor of mouth, tongue, parotid region, cheek or lip (and can extend through several regions).

An upper respiratory tract infection or trauma can result in a sudden increase in size and upper airway obstruction may occur necessitating tracheostomy. Involvement of the tongue may result in macroglossia and there is often significant cosmetic and functional disability (*Figure 6*).

Figure 4. Fourth branchial arch anomaly – previous incision and drainage of neck abscess.

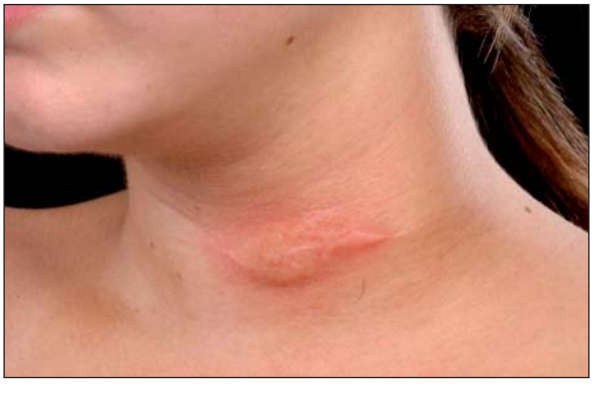
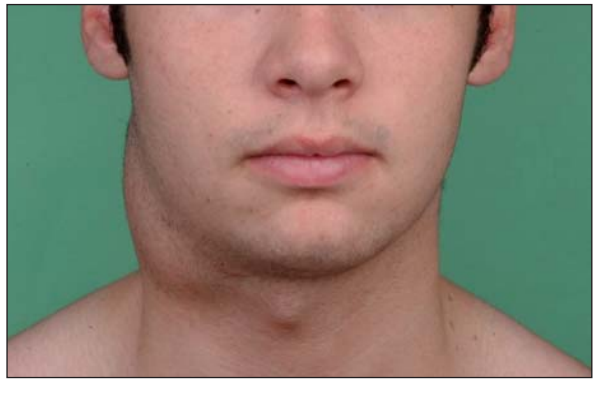


Figure 5. Branchial cyst.



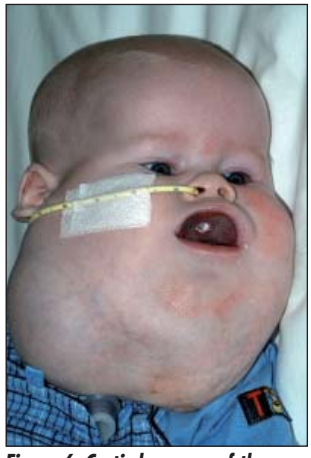


Figure 6. Cystic hygroma of the neck and tongue causing upper airway obstruction.

The diagnosis is usually clinical in combination with imaging techniques such as MRI.

Sclerotherapy under ultrasound control can be effective in 'macrocystic' lesions but surgical excision may be required. This can be challenging and is often subtotal in practice as these lesions are often closely associated with vital structures.

These children often have facial disfigurement and psychological support in addition to input from speech and language therapists and dieticians is vital.

Parotid swellings

The majority of these are infective with mumps being the commonest. Serology will confirm the diagnosis. Acute bacterial infections also occur (most commonly staphylococcal and streptococcal infection) and antibiotics may be required. On occasion incision and drainage of an abscess may be necessary. Recurrent parotitis can occur in childhood but nearly always resolves by puberty. Atypical mycobacterial infection may affect the parotid gland (see above). Branchial anomalies are mentioned above. Benign tumours such as pleomorphic adenomas are rare but are managed in a similar way to adults.

Head and neck malignancies

Fortunately, these are rare but they often present as an asymptomatic mass in the neck. Sinister symptoms include pain, dysphagia, haemoptysis, diplopia and proptosis. The incidence of head and neck malignancies in children appears to be increasing but this may be in part the result of better data collection. Fortunately, in parallel, children are now surviving these tumours in much greater numbers. Malignant tumours in children are now managed in centralized paediatric cancer units by specialized multidisciplinary teams where a number of treatment protocols have been developed and implemented.

The type of tumour varies with age and sex with, for instance, neuroblastoma the most common in infants, and thyroid carcinoma the most common malignancy in adolescent females.

Hodgkin's lymphoma

Hodgkin's lymphoma is distinguished morphologically by the presence of Reed Sternberg cells. Nodular sclerosing is the most common subtype in children and young adults. There is an association with previous Epstein–Barr virus infection. Hodgkin's lymphoma most commonly presents with lymphadenopathy in the neck (*Figure 7*).

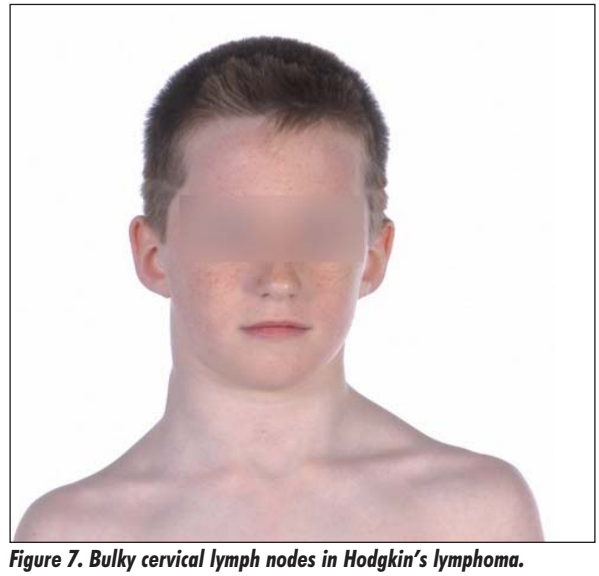


Figure 7. Bulky cervical lymph nodes in Hodgkin's lymphoma.

Systemic upsets (fever, night sweats, weight loss) are present in approximately 30% and are associated with a poorer prognosis (Hudson and Donaldson, 1997).

Diagnosis is usually made by excision of a lymph node. Investigations will include routine bloods, a chest X-ray and staging scans (combination of MRI and CT). The Ann Arbor classification is used for staging. Multimodality treatment is favoured in children and the disease-free survival is over 90% in many series.

Non-Hodgkin's lymphoma

Approximately 60% of paediatric lymphomas are non-Hodgkin's lymphoma and they tend to be more aggressive than in adults. There are three main types: lymphoblastic lymphoma (predominantly T cell), small non-cleaved cell (Burkitt's and non-Burkitt's subtype of B cell origin) and large cell (B or T cell origin). The frequency and incidence of non-Hodgkin's lymphoma varies considerably from country to country and the Epstein–Barr virus is implicated in the aetiology much more frequently in parts of Africa. Forty five per cent of children will have cervical lymphadenopathy at the time of presentation and Waldeyer's ring (including the tonsils and adenoids) is a common site of extranodal spread. Children with endemic Burkitt's often present with involvement of the jaw.

Staging investigations include relevant blood tests, bone marrow biopsy and CSF examination. CT and MRI are also used in staging (the Ann Arbor classification is used in non-Hodgkin's lymphoma). The treatment of choice is multiagent chemotherapy while radiotherapy has a limited role to play. Survival depends on the histological subtype and the extent of disease but the outlook for most children is good (Shad and Magrath, 1997).

Rhabdomyosarcoma

Forty per cent of rhabdomyosarcomas occur in the head and neck region and nearly half of these tumours present before the age of 5 years. In the head and neck region

these tumours are most frequently found in the orbit, paranasal sinuses, nose, nasopharynx and middle ear (Pappo et al, 1997). Consequently a child may present with proptosis or a mass within the nose or ear which may be associated with pain and bloody discharge (*Figure 8*). Lymph node involvement is reported to be between 3 and 36% at presentation (Coene et al, 1992).

A thorough examination of the head and neck region should be performed including the cranial nerves. An MRI scan should be performed to assess the primary lesion and to detect metastases and a CT scan is a useful complimentary tool. The Inter Group Rhabdomyosarcoma study staging system is commonly used.

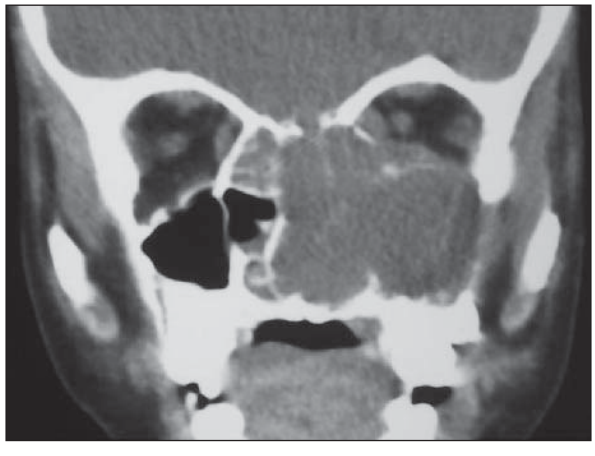
Treatment protocols have now been developed employing multiagent chemotherapy and radiotherapy. Surgery is usually performed for diagnostic biopsy only, but does have a role to play in some instances. Before 1960 only 10% of children survived 5 years. Now the 5-year survival is over 80% in early disease but still relatively poor in advanced disease and less than 10% in children with meningeal involvement.

Thyroid carcinoma

Thyroid carcinoma is rare in children (in the USA 5 cases per million per year). It is commoner in adolescents and in females. In the paediatric population 45% are differentiated papillary carcinomas, 45% are mixed papillary/follicular types and only 10% are follicular. Children usually present with an asymptomatic solitary mass in the anterior or lateral neck (cervical metastases). At presentation there is often regional lymph node involvement (74%) or distant metastases (25%). Medullary carcinoma is rare and must be suspected in children with multiple endocrine neoplasia.

Investigations will include an ultrasound scan, usually in conjunction with a fine needle aspirate biopsy. A chest X-ray, thyroid function tests and plasmathyroglobulin levels should be assessed. Plasma calcitonin levels should be measured where medullary carcinoma is suspected.

Figure 8. Computed tomography scan of an 8-year-old girl with nasal/paranasal rhabdomyosarcoma who presented with facial swelling, nasal mass and proptosis.



Treatment remains controversial because the long-term survival rates are excellent (Newman et al, 1998) and there are risks associated with radical surgery. However, in most instances, the ideal remains complete surgical excision followed by a whole-body radio-iodine scan and ablative radio-iodide therapy if necessary.

Children with a family history of multiple endocrine neoplasia should have their plasma calcitonin levels checked and should be screened for the RET proto-oncogene on chromosome 10. If positive, prophylactic total thyroidectomy is recommended (Whittemore and Cunningham, 2003).

Nasopharyngeal carcinoma

Nasopharyngeal carcinoma is uncommon in children in Europe and the United States but commoner in other locations such as parts of Africa. There is a bimodal age distribution with the first peak at the age of 10–20 years. There is an association with Epstein–Barr virus and males are twice as likely to get the disease. Children tend to have more advanced locoregional disease and distant metastases (Ayan and Altun, 1996).

Children can present with asymptomatic cervical lymphadenopathy. Others complain of nasal blockage, epistaxis, otitis media with effusion (secondary to obstruction of the Eustachian tube), headache or cranial nerve palsy (from skull base erosion). Up to 60% may have lymph node involvement at presentation. Unfortunately many of the symptoms mimic those of an upper respiratory tract infection.

Examination of the nasopharynx (usually under general anaesthetic in a child) is performed and a biopsy obtained. CT or MRI is performed to stage the disease. The American Joint Committee in Cancer staging of nasopharyngeal carcinoma is used the most.

Combined chemoradiotherapy is the treatment of choice and 5-year survival rates are similar to those of adults at 30–60% (Mertens et al, 1997). As craniofacial surgical procedures improve, surgical resection is becoming appropriate in certain situations including salvage after failed chemoradiotherapy.

Neuroblastoma

Neuroblastomas are common malignancies in early childhood and the most common malignancy in those under the age of 1 year. They arise from undifferentiated sympathetic nervous system precursor cells of neural crest origin. The adrenal gland is the most common site but tumours can arise in the sympathetic chain, posterior mediastinum and cervical regions. Lymph nodes are involved at presentation in up to 35% (Haase, 1994).

The symptoms and signs reflect the primary site and location of metastases. In the head and neck region children can present with a firm mass in the lateral neck, occasionally associated with a Horner's syndrome (cervical chain involvement). Ophthalmological mani-

festations include proptosis and periorbital ecchymosis. Bilateral periorbital haematomas are a classical sign (racoon eyes).

Examination should include a visual assessment, intraoral inspection and a thorough neurological examination. An appropriate tissue biopsy is obtained and MRI and CT imaging are usually performed. An iodine-123-metaiodobenzylguanidine (MIBG) scan is a useful method of assessing metastases. A bone marrow aspirate and trephine and urinary catecholamines should be assessed.

Treatment is tailored to the individual and the sites involved but in the neck surgical resection may be possible and may be combined with multiagent chemotherapy (Haase, 1994). Survival rates greater than 90% can be achieved in early disease. Primary neuroblastoma in the head and neck region has a better prognosis than other sites (possibly because it generally presents at an earlier stage).

Parotid malignancies

These are rare in childhood and should be suspected with a history of rapid growth of a parotid swelling, severe pain or facial weakness. Investigations will include ultrasound-guided fine needle aspirate cytology and CT or MRI scans. Wide surgical excision is required.

Miscellaneous tumours

A large variety of miscellaneous tumours can rarely affect the head and neck region and the UK Children's Cancer Study Group has formed a Rare Tumours Group which is recording data and developing some management guidelines.

A variety of sarcomas can present with pain or swelling in the head and neck region (MacGregor, 2008). There is a bimodal age distribution with incidence peaks under 5 years and in adolescents. In general, these tumours tend to recur locally and to metastasize. The treatment is usually neoadjuvant chemotherapy followed by surgery and radiotherapy as appropriate. The prognosis is usually poor.

KEY POINTS

- Swellings in the head and neck region are common in children.
- The majority are caused by reactive lymph nodes.
- Head and neck malignancies in childhood are rare and the spectrum of disease is different to that of adults.
- Excision biopsy of a lymph node remains the investigation of choice where malignancy is suspected.
- Survival rates are improving with multimodality treatment.
- Treatment of paediatric malignancies should be provided in specialized multidisciplinary paediatric oncology centres.

Squamous cell carcinomas are very rare in children and the principles of management are similar to those in adults.

Malignant teratomas in the head and neck region are rare and are often diagnosed in utero. They can compromise the airway and tracheostomy at birth may be necessary. The treatment is surgical excision (salvage chemotherapy and radiotherapy may be required).

Conclusions

A large variety of pathologies can present with a swelling in the head and neck region in children. The huge majority are benign and most commonly the result of reactive lymphadenopathy. Malignancies are rare but a number of symptoms and signs can indicate that further investigations are required. **BJHM**

Conflict of interest: none.

Ayan I, Altun M (1996) Nasopharyngeal carcinoma in children: retrospective review of 50 patients. *Int J Radiation Oncol Biol Phys* **135:** 485–92

Bloom DC, Carvalho DS, Dory C, Brewster DF, Wickersham JK, Kearns DB (2002) Imaging and surgical approach of nasal dermoids. *Int J Pediatr Otorhinolaryngol* **62:** 111–22

Coene IJ, Schouwenburg PF, Voute PA, Marion J, Burgers V, Hilgers FJ (1992) Rhabdomyosarcoma of the head and neck in children. *Clin Otolaryngol Allied Sci* **17:** 291–6

Ford GR, Balakrishnan A, Evans JN, Bailey CM (1992) Branchial cleft and pouch anomalies. *J Laryngol Otol* **118:** 19–24

Haase GM (1994) Head and neck neuroblastoma. *Semin Pediatr Surg* **3:** 194–202

Hartley B (2007) Head and neck masses. In: Graham JM, Scadding GK, Bull PD, eds. *Paediatric ENT.* Springer, New York: 112–21

Hudson MM, Donaldson SS (1997) Hodgkin's disease. *Pediatr Clin North Am* **44:** 891–906

Kubba H (2006) A child with cervical lymphadenopathy. *Clin Otolaryngol Allied Sci* **31:** 433–4

MacGregor FB (2007) Congenital cysts, sinuses and fistulae. In: Graham JM, Scadding GK, Bull PD, eds. *Paediatric ENT.* Springer, New York: 169–75

MacGregor FB (2008) Tumours of the head and neck in childhood. In: Gleeson M, ed. *Scott-Brown's Otorhinolaryngology, Head and Neck Surgery.* 7th edn. Volume 1. Hodder Arnold, London: 1251–63

Maddalozzo J, Venkatesan TK, Gupta P (2001) Complications associated with the Sistrunk procedure. *Laryngoscope* **111:** 119–23

Mertens R, Granzen B, Lassay L, Gademann G, Hess CF, Heimann G (1997) Nasopharyngeal carcinoma in childhood and adolescence. *Cancer* **80:** 951–9

Newman KD, Black T, Heller G et al (1998) Differentiated thyroid cancer: determinants of disease progression in patients < 21 years of age at diagnosis: a report from the Surgical Discipline Committee of the Children's Cancer Group. *Ann Surg* **227:** 533–41

Pappo AS, Shapiro DN, Crist WM (1997) Rhabdomyosarcoma. Biology and treatment. *Pediatr Clin North Am* **44:** 953–72

Rea PA, Hartley BE, Bailey CM (2004) Third and fourth branchial pouch anomalies. *J Laryngol Otol* **118:** 19–24

Salder TM (1985) Head and Neck. In: Tracey T, ed. *Langman's Medical Embryology.* Williams & Wilkins, Baltimore: 281–310

Shad A, Magrath I (1997) Non-Hodgkin's lymphoma. *Pediatr Clin North Am* **44:** 863–90

Whittemore KR, Cunningham MJ (2003) Malignant tumors of the head and neck. In: Bluestone CD, Stool SE, eds. *Pediatric Otolaryngology.* Saunders, Philadelphia: 1703–36