



FA Patient:

Please share this information with your ENT doctor. Bring it to every appointment.

For Ear, Nose and Throat Specialists: Head and Neck Cancer and Patients with Fanconi Anaemia

Otolaryngologists play an important role in the health care of patients with Fanconi anaemia, a rare genetic disorder. Although primarily considered a blood disease, Fanconi anaemia can affect all systems of the body and, in virtually all cases, leads to cancer.

Head and neck cancer is of particular concern for Fanconi anaemia patients. Providing a thorough examination of the patient's oral cavity at least two times per year will greatly increase the patient's overall cancer surveillance. As with all patients, early detection and treatment offer the best chance for survival.

We invite you to review the information on this flyer, and contact us directly if you have any additional questions. (Contact information is on the back of this document.) More information about Fanconi anaemia can be found on the US-based Fanconi Anemia Research Fund website, www.fanconi.org, or the UK-based Fanconi Hope website, www.fanconihope.org. Thank you!

Patients with Fanconi Anaemia (FA):

Have a 500- to 700-fold increase in the incidence of head and neck squamous cell carcinoma (HNSCC), with an increased prevalence of oral cancer

Present with cancer at a younger age than the general population, with a median age of 27 years (youngest being 10 years)

Typically present with multifocal changes, including non-cancerous lesions in addition to premalignant and invasive oral lesions

Have more aggressive HNSCC, with a two-year survival rate of less than 50% after diagnosis

Respond poorly to radiation and chemotherapy, therefore are dependent on early detection

Routine surveillance for head and neck cancer for patients with FA should:

Begin by the age of 10-12 years

Be performed on a semiannual basis by an experienced professional

Comply with the World Health Organization (WHO) oral cancer examination method (summarized on back)

Include a flexible fiberoptic exam of the nasopharynx, oropharynx, and larynx

Please turn over for information on how to conduct oral cancer examinations. For additional information, please consult the Fanconi Anemia Clinical Care Guidelines, available at www.fanconi.org or the UK FA Standards of Care Guidelines at www.fanconihope.org.

World Health Organization Standardized Oral Cancer Examination Method*

Extraoral Examination

Face: Inspect face, head, ears, and neck. Note any asymmetry or changes on the skin. Bilaterally palpate regional lymph node areas to detect any enlarged nodes.

Perioral and Intraoral Soft Tissue Examination

Lips: Observe lips with mouth both closed and open. Note color, texture, and any surface abnormalities.

Labial Mucosa: Examine the labial mucosa and sulcus of the maxillary vestibule and frenum and mandibular vestibule. Note color, texture, and any swelling or other abnormalities of the vestibular mucosa and gingiva.

Buccal Mucosa: Examine right and left buccal mucosa from the labial commissure to the anterior tonsillar pillar. Note any change in pigmentation, color, texture, mobility, and other abnormalities.

Gingiva: Examine the buccal and labial aspects of the gingival and alveolar ridges from the right maxillary posterior gingival and alveolar ridge, around the arch to the left posterior area, and reverse.

Examine the palatal and lingual aspects from right to left on the palatal and left to right on the lingual.

Tongue: With tongue at rest and mouth partially open, examine dorsum for swelling, ulceration, coating or variation in size, color or texture. Note any change in pattern of papillae covering on tongue surface and examine tip of tongue.

With tongue protruded, note any abnormality of mobility or positioning.

Using mouth mirrors, inspect right and left lateral margins of tongue.

Grasp tip of tongue and examine posterior aspects of tongue's lateral borders.

Examine ventral surface.

Palpate tongue to detect growths.

Floor: With tongue elevated, inspect floor of mouth for changes in color, texture, swellings or other surface abnormalities.

Palate: Inspect hard and soft palate with mouth wide open, head back, and tongue depressed.

Examine all soft palate and oropharyngeal tissues.

Bimanually palpate floor of mouth for any abnormalities.

Palpate all mucosal or facial tissues that appear abnormal.