

# AIRWAY EVALUATION FOR MAGNETIC RESONANCE IMAGING SEDATION IN PEDIATRIC PATIENTS WITH PLEXIFORM NEUROFIBROMA

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## Introduction

Neurofibromatosis type 1 (NF1) or Von Recklinghausen disease, the most common form of NF, is an autosomal dominant disease with a variable expressivity and a wide variety of clinical manifestations. In one half of cases, NF-1 can result from a de novo mutation, with no previous family history of disease. It affects males and females equally with a disorder frequency of 1 in 4,000<sup>1</sup>. The gene affected in NF-1, is located on the long arm of the chromosome 17 (q11.2). Neurofibromatosis modifies neurofibromin, a "tumor suppressor" protein, allowing rapid growth of cells, especially around the nervous system. This leads to the common symptoms of neurofibromatosis. The incidence of head and neck involvement in patients with NF varies between 14% and 37%<sup>2</sup>. Plexiform neurofibroma of the neck is a cause of morbidity in the affected individual with possible airway abnormalities. Challenges in the care of these patients include the evaluation and the determination of airway patency prior to anesthesia/sedation for the MRI exam.

## Background

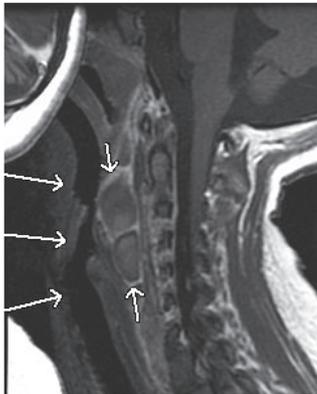
Pediatric patients with NF 1 may present for sedation for MRI exam as outpatients with a growing large neck mass. Previous MRI exams may suggest involvement of the airway with the progression of the tumor. Picture 1 shows an example of a neck MRI of a large plexiform neurofibroma involving predominantly the left side of the neck. The lesion is seen in the left carotid space, extending through the left lateral neck into the supraclavicular fossa to the brachial plexus and then into the intercostal space and dorsal paraspinous region. The lesion pushes the parapharyngeal space forward and seems to compress the airway. Verifying the patency of the airway prior to proceeding in anesthetizing patients with evolving face/neck masses may require additional interventions, such as a neck and chest X ray, an otolaryngology consult prior to sedation, a flexible/rigid fiberoptic laryngoscopy, or a CT scan exam. The pediatric patient offers a unique challenge in the risk of exposure to radiation and the need of sedation in order to insure immobilization during different type of examinations.

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Picture 1

The long arrow points to the compressed hypopharyngeal airway, the short arrow points to the superior aspect of the left pyriform sinus. The right pyriform sinus is not seen.



## Discussion

Preoperative assessment of the airway is focused primarily at the detection and evaluation of laryngotracheal obstruction and involvement of adjacent structures. A postero-anterior and lateral neck and chest radiographs may be sufficient in some patients to assess the degree of tracheal compression and deviation; however, more modern imaging techniques such as a magnetic resonance imaging (MRI) provide more detailed information about the extent of airway involvement and the degree of mass extension. MRI can produce imaging of the airway without incurring a large dose of radiation to the patient; these images are susceptible to artifact from movement and therefore require sedation of the pediatric patient. Sedation may aggravate airway narrowing secondary to collapse of the tissues, therefore in the case of suspicion of airway involvement; a detailed assessment prior to sedation is requested.

Computerized Tomography (CT) scanning may offer an alternative way for airway assessment because of fast imaging but with the risk of exposure to radiation. There are limitations of axial CT images for assessing the airways such as limited ability to detect subtle airway stenosis and craniocaudal extent of disease; difficulty displaying relationships of the airway to adjacent mediastinal structures; inadequate representation of airways oriented obliquely to the axial plane; and difficulty assessing the interfaces and surfaces of airways parallel to the axial plane<sup>3</sup>.

Complementary ways of viewing the data from the original axial CT data set can help to overcome these limitations. The CT data can be reconstructed into two-dimensional (2-D) reformations and three-dimensional (3-D) images, including internal virtual endoscopic (VE) renderings that simulate images from conventional bronchoscopy<sup>4</sup>. Detection of lesions in the airway using virtual bronchoscopy was reported to reach a high sensitivity (>90%) for lesions that were > 5 mm in diameter. However, there was limitation by a high false-positive rate due to the difficulty in differentiating retained secretions from the airway<sup>5,6</sup>. External 3-D rendering of the airways, or CT tracheobronchography, depicts the external surface of the airway and its relationship to adjacent structures. Some authors suggested that VE may be considered as a substitute to direct endoscopic examination sparing them an extra anesthetic for evaluation<sup>7</sup>. However, VE may have some limitations and does not provide histology, and it cannot identify functional lesions of the vocal cords<sup>8</sup>. In the pediatric population sedation/anesthesia may be needed in order to perform the CT scan and adequate justification is required so that the perceived benefits outweigh the risks of using ionizing radiation. Flexible bronchoscopy and CT scan are considered complementary techniques in the evaluation of laryngeal function and during follow-up while rigid bronchoscopy remains the procedure of choice in the evaluation of candidates for tracheal resection and reconstruction for postintubation stenosis<sup>9</sup>. Although, the 3-D rendering of CT scanning in the pediatric population<sup>4,10</sup> may be considered as a useful adjunctive radiological tool in airway assessment; MRI does not involve ionizing radiation and is valuable in demonstrating the relation of the airway to adjacent blood vessels without injection of intravascular contrast. MRI may be considered as the preferred modality for assessing paratracheal abnormalities in children<sup>11</sup>.

Successful management of plexiform neurofibromas of the head and neck in patients with neurofibromatosis type 1 (NF1) requires detailed preoperative planning. Magnetic resonance imaging (MRI) is indicated preoperatively to avoid the associated loss of function and to delineate precisely the extent of the tumor. Challenges in the care of these patients include the evaluation and the determination

of airway patency prior to sedation for the MRI exam. Although, computed tomography can yield useful information, the most definitive technique for upper-airway evaluation involves the direct visualization of the anatomy and dynamics<sup>12</sup>. Because of the specific advantages and limitations of each technique, the combined use of flexible and rigid endoscopes, with a carefully planned approach to sedation and anesthesia, yield to the most accurate diagnostic information.

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