# Management Of The Difficult Airway In A Patient With Tuberous Sclerosis: A Case Report

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# Abstract

During surgical procedures of head and neck lesions, management of the airway is always a problem and anticipation of difficulties in intubation have to observed, alternative maneuvers for intubation may be necessary. We present the case of an 18-year-old male with tuberous sclerosis who required intubation because of facial deformity secondary to progressive tumor growth and debunking was planned, modifications to classic maneuvers are discussed. We hope this case presentation will be valuable in increasing the awareness of physicians about this rare cause or difficult intubation, and to have in mind alternative maneuvers basically when limited resources are the main obstacle.

# BACKGROUND

One of the most important issues and concerns during surgical procedures of head and neck lesions is the problematic of management of the airway, defining difficult airway as the clinical situation in which there exists a difficulty for ventilation with mask, difficulty for endotracheal intubation, or both, and difficult intubation, such as endotracheal catheter placement that requires more than three attempts or more than 10 minutes to perform intubation <sup>(1)</sup>. Other definitions cite difficult airway as the following: more than two attempts with the same blade; change of blade or an adjunct blade for direct laryngoscopy, and the use of a later alternative technique after failed intubation<sup>(2)</sup>.

Prior appraisal is of highest importance because it aids in anticipating when the airway is difficult to approach, for which different scales have been described, such as those of Mallampati, Patil, Bellhouse and Dore, and Comak, among others<sup>(3-5)</sup>. However, there are very particular cases in which these scales are not helpful because soft tissue lesions of the head and neck region will cause intubation to be difficult in itself.

Distinct maneuvers that facilitate visualization of the larynx have been described: a) Posterior displacement of the larynx by putting backward pressure on the thyroid or cricoids cartilage (BACK)<sup>(4,6)</sup>; b) Backward upward rightward pressure (BURP)<sup>(6,7)</sup>; c) Optimal external laryngeal manipulation (OELM)<sup>(8)</sup>; d) bimanual manipulation<sup>(9)</sup>, and e) mandibular advancement<sup>(10)</sup>, as well as distinct positionings of the head to facilitate exposure of laryngeal structures, such as sniffing position and head elevated laryngoscopy positioning (HELP)<sup>(10-12)</sup>.

Combinations of maneuvers have been recommended, including head elevation and external laryngeal pressure to improve laryngeal visualization<sup>(11,12)</sup>, BURP maneuver, and mandibular advancement, which are frequently helpful in fiber optics-enhanced intubation<sup>(10)</sup>.

In the present case, we show an example of the approach to the difficult airway in a patient with a large tumor of the facial soft tissues, and we advance a proposal for the management of patients found under similar conditions or in settings where technological resources are the limitations.

# **CASE REPORT**

A 19 year old Mexican male with the diagnosis of tuberous sclerosis at the age of 8 years presented with multiple small cutaneous tumors, some pendunculated and others sessile, violet in color of bland consistency and vascular aspect, localization predominantly in left side of the face, as well as with a subcutaneous tumor in left maxillary region of progressive growth to complete deformation, occluding the left eye by over 70%, displacing the dental arch, the nasal pyramid, and extending to the ipsilateral preauricular region. The patient had been administered treatment with radiotherapy to this site in 2006 without tumor shrinkage of

the tumor. It was decided to program the performance of left lateral rhinotomy and left maxillectomy in May 2007, prior to Magnetic resonance imaging (MRI) with reconstruction (Figures 1 and 2).

## Figure 1

Figure 1- Facial deformity and location of larynx.



# Figure 2

Figure 2- Magnetic resonance reconstruction evidencing involvement of the maxilla .



Pre-anesthetic evaluation scores for difficult airway were as follows: Mallampati, IV; Patil, 3; Interincisive distance (ICD), III, and Bellhouse Dore, I. From the patient's arrival at the operating room, he was monitored with continuous Electrocardiography (EKG), T/A, and pulse oximetry, and equipment was prepared to include endotracheal catheters, guides, fibrolaryngoscopy, aspiration catheter, and adjunct anesthesiology.

Previous oxygenation with mask and with the patient conscious under sedation with midazolam 2 mg, as well as fentanyl 200 lg; direct laryngoscopy was performed by means of OELM and armed endotracheal catheter placement (Figure 3).

# Figure 3

Figure 3- Head positioning, rigid laryngoscope movement aided by arm movement and laryngeal displacement (Arrows).



During the surgical procedure, the patient was hemodynamically stable with adequate ventilatory parameters (Figure 4).

#### Figure 4

Figure 4- Successful intubation and placement during surgery.



At the end of surgery, extubation was conducted with the patient awake, without complications. Postoperatory evolution was adequate; thus, the patient was discharged from the hospital 3 days after the surgical procedure, has been followed-up to these days without complications or recurrence of the facial tumor.

# DISCUSSION

Adequate appraisal of the patient is necessary, because it aids in anticipating difficult airway. There are simple and non-invasive scales for evaluation for suspicion of the condition, including the following: the Mallampati instrument, which visualizes the pharyngeal structures when the patient has his/her mouth open, with four types<sup>(3)</sup>; the Patil, which considers the thyromental distance with neck in hyperextension with three classes; Interincisive distance describes four degrees (I, >3 cm; II, 2.5–3 cm; III, 2–2.5 cm, and IV, <2 cm), and Bellhouse and Dore, which evaluates the degree of extension of the head, with four types<sup>(4)</sup>. Cormack described visualization of laryngeal structures during direct laryngoscopy, subdividing these into four stages<sup>(5)</sup>. Some authors have subdivided visualization into three degrees: good visualization (glottic ring is visualized); poor visualization (solely the arytenoids are visualized), and non-visualization (only the epiglottis is visualized)<sup>(13)</sup>. These scales possess high sensitivity, but low specificity and low predictive value; thus, maneuvers for facilitating laryngeal visualization and with this, intubation, are important.

The following are recommended within the management guides for difficult airway approach (clinical indications):

- Adequate positioning for intubation, head extension, and neck flexion (sniffing position), as well as necessary measures for difficult airway approach, catheters, guides, laryngeal blades, etc.
- 2. Secondary tracheal intubation, utilization of ventilatory devices such as fiberscope, fast track, laryngeal masks, laryngeal tubes, etc.(14-16).

With the purpose of facilitating intubation, diverse maneuvers have been designed to facilitate visualization of the larynx, which are described as follows: the BACK maneuver, which consists of posterior displacement of the larynx, pressuring cricoid cartilage backward, improving visualization of laryngeal structures with reduction of the incidence of failure to visualize some portion of the epiglottis in degrees IV and V of  $8.0-3.0\%^{(4,13)}$ ; the BURP maneuver with thyroid-cartilage larynx displacement in three specific directions, later toward the cervical vertebrae, toward the upper part to the greatest degree possible, slightly lateralizing rightward, which results in better visualization of the epiglottis which diminishes the intubation failure rate<sup>(4,13)</sup>, and external laryngeal manipulation, also termed bimanual manipulation, which exerts cricoid or BURP-type pressure, performed initially with the right hand of the Laryngoscopist and later maintained by an assistant. Improved visualization of laryngeal structures has been shown, and laryngeal approach was permitted <sup>(6,7,16)</sup>.

Positioning of the head has also been cited as determinant during airway approach; diverse authors, such as H. Schmitt, R. Levitan, and S. Shivanna, have published that head elevation and neck flexion significantly improve visualization of the epiglottis, noting that elevation of the head causes a later movement of the epiglottis, as well as relaxation of the frontal muscles of the neck, which allows for great exposure of the larynx, this also termed Head elevated laryngoscopy positioning (HELP)<sup>(11,12,17)</sup>. In addition, the combination of head elevation and external laryngeal pressure for improving laryngeal visualization has been recommended, exhibiting improvement of visualization from grade 3 to grade 22<sup>(11,12)</sup>. All of these entertain the purpose of endotracheal intubation; despite all of this, there is the possibility of not being able to intubate the patient. Thus, it is necessary to improve this external optimal laryngeal manipulation. We described herein the approach for accessing the airway in a patient with a diagnosis of tuberous sclerosis and maxillary tumor in left hemiface with extensive deformity that encompasses nasal septum and

mouth.

At National Institute of Cancerology in Mexico City difficult airway patients is a very common finding, bimanual manipulation or optimal external laryngeal pressure recommended by certain authors at the thyroid and cricoid cartilage level<sup>(7-9)</sup> and cited by some authors for the area of the neck, as well as required pressure, cannot be performed because of the large tumors that we have. In situations like these, we recommend external laryngeal manipulation below the described level, specifically, at the supraexternal notch which permits better displacement of the laryngeal-tracheal body in cephalic direction, allowing better laryngealstructure visualization, which in combination with the HELP position with >10-cm head elevation, facilitates maximal neck flexion, permitting relaxation of all frontal muscles of the neck, improving exposure of the laryngeal structures  $^{(11,12,17)}\!.$  This combination has been found to be the best combination of maneuvers facilitating intubation in less time and with a lesser number of attempts in the performance of successful intubation.

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