

## Airway Management in Stickler Syndrome Patient: A Case Report

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Received date: October 07, 2017; Accepted date: October 27, 2017; Published date: October 30, 2017

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### Abstract

**Background and objectives:** Stickler syndrome is a recognized syndrome by its distinctive craniofacial dysmorphism. It is a rare medical condition caused by collagen alpha chain mutations, with subsequent conjunctive tissue disruption and multisystem involvement. We present a clinical report of an expected difficult airway management in a Stickler syndrome female patient.

**Case report:** Stickler's syndrome patient scheduled for left mastoidectomy and tympanoplasty under general anesthesia. Expected difficult airway. Induction and tracheal intubation was performed without uneventful.

**Discussion:** Expert airway management is an essential skill in anesthetic practice. The case highlights the importance of preoperative assessment of the personal and familiar background and the physical examination in order to planning difficult airway management.

**Keywords:** Stickler syndrome; Airway management

### Introduction

Stickler syndrome, also known as hereditary progressive arthropathy [1], is a recognized syndrome by its distinctive craniofacial dysmorphism affecting 1 in 7,500 to 9,000 newborns [2]. It is a rare medical condition caused by collagen alpha chain mutations, with subsequent conjunctive tissue disruption and multisystem involvement. Most patients have autosomal dominant transmission; some autosomal recessive cases have been described [2]. Individuals with Stickler syndrome may have flattened facial appearance, short nose, anteverted nostrils, ocular findings of myopia, cataract, retinal detachment, cleft palate, mitral valve prolapse of the heart, hearing loss that is both conductive and sensorineural and double jointedness [1,2]. These patients can also develop spondyloepiphyseal dysplasia and precocious arthritis [2]. Variable phenotypic expression of this syndrome occurs within families [2].

As patients may have multiple surgeries due to this syndrome's several complications, an anesthesiologist timely intervention is critical. To our knowledge, there is no available data regarding anesthetic airway management in Stickler's syndrome patients. Cook et al. in the 4<sup>th</sup> National Audit Project of the Royal College of Anaesthetists (NAP4) observed the incidence of one serious airway complications per 22000 general anesthetics and the expected difficult intubation was recorded in 42% of the cases [3].

We present a clinical report of an expected difficult airway management in a Stickler syndrome female patient.

### Case Report

A Stickler's syndrome 33 years-old female, American Society of Anesthesiologists (ASA) physical status II was scheduled to left mastoidectomy and tympanoplasty under general anesthesia. Clinical past history related to the syndrome included generalized arthropathy due to congenital spondyloepiphyseal dysplasia, cleft palate with surgical intervention in childhood and deafness. There were no available anesthetic medical records concerning previous bilateral total hip replacement procedures, although the patient's report that the procedure was performed under general anesthesia, without interurrences. Regarding familiar background, Stickler syndrome was present in several relatives and the patient described the occurrence of an emergency cricothyroidotomy during a c-section on patient's mother.

Preanesthetic evaluation revealed an expected difficult airway as we show in figure 1 with Mallampati class II, ogival palate, cervical mobility <90 degrees, prominent superior incisors with >4 cm mouth opening, tiromentonian distance >6 cm and upper lip test class II. It was explained to the patient the risk of difficult airway and the anesthetic team advertised her about the orotracheal tube removal only after she was fully awake and complete reversal of airway reflexes.

The difficult airway devices such as bougies, aintree intubation catheter, airway exchange catheter, short handle laryngoscope, McCoy blade, straight blade, supraglottic devices, videolaryngoscope and flexible intubating fiberoptic bronchoscopy were available in the operating room.

After standard ASA, Bispectral Index and Train-Of-Four monitoring, preoxygenation was performed using 10 H<sub>2</sub>O continuous positive pressure in a semisitting position. A remifentanil 0.15 mg/kg/min infusion was started and a 2 mg/kg bolus of propofol was

administered. Facemask ventilation was successfully achieved and rocuronium 0.6 mg/kg was given to optimize the conditions of orotracheal intubation.

Direct laryngoscopy revealed a Cormack-Lehane classification grade 3 with back-up right pressure and tracheal intubation was performed with RAE (Ring, Adair, Elwyn) endotracheal 7 mm tube with stylet and metallic McCoy blade. The anesthesia was maintained with sevoflurane to adequate BIS values and remifentanil infusion.

Patient remained hemodynamically stable intraoperatively. Undesirable events did not occur during surgery. When the procedure ended, TOF was 33% and BIS 45. It was given 2 mg/kg of sugammadex with blockade reversion in 75 s (TOF 92%). The halogenated was stopped and the patient had awakened about 5 min later. She fulfilled orders and the extubation was performed at the end of a deep inspiration without any undesirable events.

In the postoperative period, a difficult airway letter was delivered to the patient. Figure 1.



**Figure 1:** Physical examination of the stickler's syndrome female.

## Discussion

Stickler syndrome is a clinical situation with multiple phenotypic expression. The medical complications depends on the syndrome's evolution, so these patients are potential surgery candidates.

The peri-operative morbimortality is potentially not negligible although there are no case reports in the literature. A thorough preoperative assessment of the personal and familiar background and the physical examination allowed us the recognition of a potentially difficult case. Individuals usually present physical abnormalities on airway examination and cardiac manifestations related to Stickler syndrome could be an issue of concern. Because of that, it was made an early and adequate anesthetic planning, which revealed to be the cornerstone to the success of this case.

In the preanesthetic evaluation, the authors observed an extremely anxious patient not cooperative for an awake intubation technique. Due to the absence of any interferences related to past bilateral hip arthroplasty under general anesthesia and the anesthesiologist extensive experience in difficult airway, the anesthetic team decided to

manage the airway under general anesthesia with the following safety approach. During preoxygenation an adequate capnography curve was checked with an end tidal of O<sub>2</sub> 92%, which indicated facial mask's optimal adaptation and viability of effective ventilation. These parameters were observed and classified as non-emergent airway. Furthermore, the drugs used in anesthetic induction have a rapid washout and their effects are rapidly reversed.

Remifentanil is a drug with a short half-life and with the possibility of rapid reversal of the effect with naloxone, so it is useful in the management of a difficult airway. Moreover, its anxiolytic action allowed to better performing the preoperative preparation in a patient who was already very anxious. Regarding the surgical procedure (tympanoplasty and mastoidectomy), the improvement of the surgical field conditions, with less possible hemorrhage, was thus better achieved.

The anesthetic team decided to administer rocuronium to optimize the conditions of orotracheal intubation, only after checking ventilation with facial mask was performed without difficult. This administration was performed with difficult airway material available and 16 mg/kg dosis of sugammadex in the operating room (despite we consider that the sugammadex accessibility should not be a pretext for indiscriminate use of rocuronium). On the other hand, surgeons and the material were available if invasive techniques were needed. The curameter was used during the entire anesthetic procedure: it was turned on after anesthetic induction to obtain supramaximal response and it was maintained until complete reversal of muscular block. It is a common practice in our institution this monitoring in case of muscular relaxants use.

Although the authors recognize that videolaryngoscopy use would be another acceptable option and it was available in the operating room, direct laryngoscopy with McCoy blade was first performed by an anesthesiologist experienced in difficult airway management using conventional laryngoscopy and orotracheal intubation was achieved with stylet without complications. Approach used allowed the anesthesiologist to document the grade of direct laryngoscopy for possible future interventions

In the authors opinion more publications related to anesthetic procedures in patients with this syndrome are needed to improve the anesthetic care.

The preanesthetic assessment and the anesthetic planning are crucial and can contribute to decrease perioperative adverse events.

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