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General Anesthesia for Dental Procedures in Patients with Fibrodysplasia Ossificans Progressiva: A Review of 42 Cases in 30 Patients

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Abstract

Background—Fibrodysplasia ossificans progressiva (FOP) is a rare genetic condition characterized by progressive heterotopic ossification of skeletal muscle and soft connective tissues, leading to progressive ankylosis of all joints of the axial and appendicular skeleton. Cervical spine fusion, ankylosis of the temporomandibular joints, thoracic insufficiency syndrome, restrictive chest wall disease, and sensitivity to oral trauma complicate airway management and anesthesia and pose life-threatening risks.

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Methods—We conducted a retrospective chart review at one institution of patients with FOP who underwent general anesthesia (GA) for dental procedures.

Results—Thirty patients underwent 42 general anesthetics. In 35 of 42 cases, GA was induced after the airway was secured by an awake fiberoptic intubation. In 4 of 42 cases, all of them pediatric, GA was first induced with maintenance of spontaneous ventilation, and the trachea was then intubated using a fiberoptic scope. In two cases, one adult and one pediatric, GA was first induced, and the trachea was then intubated using a GlideScope®. In one case, the patient had a cuffed tracheostomy device in place that was accessed for GA. In 36 of 42 cases, the patients were discharged to home on the same day as their dental procedure. No significant postoperative complications were encountered.

Conclusions—GA can be administered safely to patients with FOP for dental procedures with attention to perioperative and airway management using a multidisciplinary approach. An awake nasal fiberoptic intubation should be considered the first choice for airway management. Most patients can be discharged to home on the same day as their dental procedure.

Introduction

Fibrodysplasia ossificans progressiva (FOP) is a rare, progressive, and disabling autosomal dominant disorder of extraskeletal endochondral ossification.¹ The diaphragm, tongue, extraocular, and laryngeal muscles are spared from heterotopic ossification; cardiac and smooth muscles are also not affected.² Individuals with FOP appear normal at birth except for malformations of the great toes. During the first decade of life, inflammatory soft tissue swellings, also known as flare-ups, progressively transform skeletal muscles and connective tissues into a second skeleton of heterotopic bone that progressively immobilizes all of the joints of the axial and appendicular skeleton.² Flare-ups may occur spontaneously, but can be precipitated by misplaced venipuncture, soft tissue injury, muscle fatigue, intramuscular injection, biopsy or excision of heterotopic bone, viral illnesses, routine dental therapy, and injection of local anesthetics during dental procedures.²⁻⁴

Disease flare-ups are episodic; immobility is cumulative. Heterotopic ossification usually begins by ten years of age with neck and shoulder involvement.^{1, 2, 5} The temporomandibular joints may be involved early and are vulnerable to trauma at any age.³ The median lifespan is 40 years; death results most commonly from complications of thoracic insufficiency syndrome or pneumonia.^{6, 7} A few deaths have been attributed to complications of general anesthesia (GA).^{6, 7} At the present time, no medical or surgical intervention can alter the natural progression of FOP.⁸

Anesthetic management for patients with FOP is challenging. Cervical spine fusion, ankylosis of the temporomandibular joints, thoracic insufficiency syndrome, restrictive chest wall disease, and sensitivity to oral trauma complicate airway management and anesthesia and pose life-threatening risks. Although several single-patient cases of the anesthetic management of patients with FOP have been reported,⁹⁻²² no one institution has reported on a large series of FOP patients. The Departments of Anesthesiology and Dentistry at Thomas Jefferson University Hospital (TJUH) and the Center for Research in FOP and Related Disorders at the University of Pennsylvania collaborate in the multidisciplinary care of FOP

patients. Many patients with FOP travel to these institutions for their medical and dental care. The anesthesia providers at TJUH as a result have managed many patients with this rare disease.

We performed this review to evaluate 1) the safety of anesthetic and airway management in patients with FOP undergoing dental rehabilitation and 2) the ability to discharge these patients to home on the same day of surgery. In this case series, we reviewed the perioperative courses of 30 FOP patients involved in 42 cases.

Methods

After receiving approval from the IRB of TJUH, we reviewed the charts of patients with FOP who had GA for dental procedures from December 2001 through December 2011. The patients' paper charts, electronic anesthesia preoperative and intraoperative charts (after June 2006), and electronic hospital clinical charts were reviewed. The IRB waived the requirement of written informed patient consent.

For each anesthetic, the following were recorded: the patient's age, gender, weight, height, ASA physical status classification, Mallampati classification and cervical mobility (both determined by the anesthesiologist who performed the preoperative evaluation), anesthetic history, medical and surgical histories. Additionally documented were: the anesthetic techniques for intubation of the trachea and induction of GA, sedation and induction drugs, maintenance drugs, corticosteroid administrations, duration of time in the operating room before the start of dental surgery, duration of surgery, recovery time, preoperative admissions, overnight admissions, and postoperative complications. Involvement of medical disciplines other than anesthesiology or dentistry was evaluated. Statistical data analysis (mean, median, standard deviation, and range) was performed using Microsoft Excel®.

Results

Thirty patients with FOP underwent 42 GAs for dental rehabilitation. Seven patients had multiple anesthetics. The mean age at the time of surgery was 23 $\pm$ 14 years (median 20, range 5 to 64 years). Females comprised 57% of the patients and 60% of the anesthetics. The mean weight of the patients was 53 kg $\pm$ 24 (median 49, range 11 to 118 kg); the mean height was 158 cm $\pm$ 19 (median 160, range 91 to 183 cm); and the mean Body Mass Index was 20 kg/m² $\pm$ 6 (median 19, range 9 to 37 kg/m²). The Mallampati classification scores were: 15% (6 of 40) class I, 5% (2 of 40) class II, 7.5% (3 of 40) class III, and 72.5% (29 of 40) class IV (two scores were not documented). The patients' ASA physical status classifications at the time of their anesthetics were: 14% (6 of 42) ASA physical status II, 79% (33 of 42) ASA III, and 7% (3 of 42) ASA IV.

GA was administered to all of the patients for their dental rehabilitation procedures. The technique used most frequently (35 of 42 cases) was GA after an awake fiberoptic intubation. All of the awake fiberoptic intubations except one were performed with the patient sedated. One of these intubations was oral; the rest were nasotracheal.

This case series includes 19 pediatric anesthetics (patients younger than 18 years old) in 17 patients. In 14 of the 19 pediatric cases, a sedated fiberoptic approach to airway management was used. In 4 of the 19 pediatric cases, GA was first induced with maintenance of spontaneous ventilation, and the trachea was then intubated through the nasopharynx using a fiberoptic scope. The ages of these patients were 5, 7, 9, and 10 years old. Their airway examinations respectively were: class III with no cervical motion, class IV with full cervical motion, class I with full cervical motion, and class IV with undocumented cervical motion. In one pediatric case involving another 9-year-old patient, GA was first induced with maintenance of spontaneous ventilation, and the trachea was then nasally intubated using a GlideScope®. This patient's airway examination was a class I with limited neck extension. Care was taken not to overextend the temporomandibular joints of this patient; the range of mouth opening was measured before and during intubation. No difficulty with ventilation was noted in any of the pediatric cases where GA was induced before securing the airway.

In all but 2 of the adult cases, the airway was secured using an awake, nasotracheal fiberoptic intubation technique followed by induction of GA. In one case, GA was first induced, and the trachea was then nasally intubated using a GlideScope®. This patient had a Mallampati class I score and full range of cervical motion. The range of mouth opening was measured before intubation; the jaw was not hyperextended during intubation. In the other case, the patient had a chronic, cuffed tracheostomy device in place. The device was connected to the anesthesia machine, and an inhaled induction of GA was performed.

Practitioners from numerous medical specialties were involved in patient care. In 30 cases, the patient was evaluated by an anesthesiologist before the day of the procedure in the preoperative testing center. In nine cases, the patient was first seen by an anesthesiologist in the operative holding area; two of these patients had been evaluated before a past anesthetic. In three cases, the patients were admitted to the hospital preoperatively and evaluated by an anesthesiologist in the hospital. An otolaryngologist was immediately available during every case to assist with intubation or perform an emergency tracheostomy if needed. An otolaryngologist electively assisted the anesthesiologist with a fiberoptic intubation in five cases. In one case, an otolaryngologist evaluated a patient's bronchial tree after a successful fiberoptic intubation by anesthesia (this patient was diagnosed with narrowing of the left mainstem bronchus due to anterior displacement of the posterior tracheal wall). A dentist primarily performed all of the dental rehabilitation procedures, but an oral and maxillofacial surgeon assisted the dentist by performing tooth extractions in 34 of the cases. A cardiologist was consulted preoperatively in two cases, and a family medicine physician was consulted preoperatively once. A pediatrician admitted the patient to the hospital preoperatively in two cases and postoperatively in two cases. In one case, an intensivist admitted the patient to the intensive care unit postoperatively.

The mean duration of time in the operating room before the start of dental surgery was 54 +/- 22 minutes (median 52 minutes, range 21 to 120 minutes). Neuromuscular blocking drugs were never administered. Various drugs were administered for GA maintenance; sevoflurane was used most frequently (40 of 42 cases). In 31 of 42 cases, the patient received intraoperative corticosteroids.

The mean duration of surgery was 125 $\pm$ 76 minutes (median 116 minutes, range 11 to 315 minutes). All of the patients were tracheally extubated in the operating room. The average time spent in the postanesthesia care unit was 70 $\pm$ 25 minutes (median 67 minutes, range 28 to 150 minutes); one patient went directly to the intensive care unit due to worsening of his chronic hypoxemia.

Thirty-six of the 42 patients were discharged from the hospital on the same day as their procedures; six were admitted overnight. The reasons for the admissions were: brittle diabetes, a history of malignant hyperthermia, worsened hypoxemia after extubation that required close monitoring, threat of airway swelling due to a newly drained abscess, multiple comorbidities (diabetes, asthma, restrictive lung disease, hypothyroidism, and history of a difficult airway and tracheostomy), and an undocumented reason. One patient complained of severe shoulder pain in the recovery room; his surgical time was the longest at 315 minutes. His pain resolved within a few days, and no neuropathy developed.

Discussion

This large case series demonstrates that GA for dental procedures can be safely accomplished in patients with FOP using a multidisciplinary approach. No major anesthetic complications occurred in our study. Although a cannot ventilate/cannot intubate situation was not encountered in this case series, three such cases have been reported.^{9,10,11} Because of the difficult airway management reports in the literature and our use of numerous medical specialties, we recommend that patients with FOP are cared for at an institution where a multidisciplinary approach is possible. An anesthesiologist should evaluate the patient preoperatively, preferably before the day of the procedure. An otolaryngologist should be immediately available during the procedure to assist with airway management and perform an emergency tracheostomy if needed. A dentist and an oral maxillofacial surgeon should be involved in each case so that a comprehensive oral rehabilitation with tooth extractions can occur under one GA, which is safer and more convenient for the patient. Other medical practitioners, such as pediatricians, family medicine physicians, cardiologists, and intensivists, may be required.

This case series demonstrates that most FOP patients can be discharged to home on the same day as their dental procedures. This finding has not been previously reported. Only one case report in the literature involved a dental procedure; that patient was discharged from the hospital on postoperative day four, after “confirming the absence of medical or nutritional problems.”²¹

Limitations of this case series are due to the series’ retrospective nature. Reasoning for detailed intraoperative management decisions, such as the decision for or against an awake fiberoptic intubation, was not always clear from the information provided in the chart and discussion with the anesthesia providers. Some desired information was not recorded in every patient.

In two cases, the patients’ tracheas were intubated using a GlideScope[®] after induction of GA. Both patients had normal mouth opening. Jaw overstretching may cause tissue trauma

and subsequent flare-up.³ Several incidents of temporomandibular joint flare-ups after jaw overstretching have been reported to one of the authors (FSK). Intubation using a GlideScope® and direct laryngoscopy with a blade should be avoided in FOP patients with decreased mouth opening. We believe that the nasotracheal approach to intubation in FOP patients lessens the need for jaw overstretching, thus reducing the risk for postoperative temporomandibular joint flare-ups. Also, laryngoscopy using a Glidescope® or a direct technique in an FOP patient with decreased mouth opening may be difficult.

An awake nasal fiberoptic intubation should be considered the first choice for airway management. This recommendation is based on the potential for difficult airway management and the risk of jaw overstretching associated with a direct laryngoscopy. We acknowledge, however, that induction of GA before securing the airway may be appropriate in an extremely uncooperative or anxious pediatric patient. The 2013 “Practice Guidelines for Management of the Difficult Airway” communicate, “The uncooperative or pediatric patient may restrict the options for difficult airway management, particularly options that involve awake intubation. Airway management in the uncooperative or pediatric patient may require an approach (e.g., intubation attempts after induction of general anesthesia) that might not be regarded as a primary approach in a cooperative patient.”²³

There were several reasons for choosing GA with an endotracheal tube in this case series. First, routine injections of local anesthetic for dental procedures, especially mandibular blocks, should not be used because they can precipitate flare-ups and cause fusion of the temporomandibular joints.³ Second, oral access can be difficult if a patient has decreased mouth opening. If a tooth fragment were dropped in the mouth of an unintubated patient with a fused jaw, it could be impossible to retrieve. An endotracheal tube provides needed airway protection. Third, the dentist and the oral and maxillofacial surgeon may need Trendelenburg positioning for long periods of time. This positioning may cause patient discomfort and result in respiratory compromise, especially in patients with preexisting pulmonary disease.

One of the patients in this series complained of severe shoulder pain in the recovery room. Inadequate positioning and extended length of the procedure (315 minutes) may have been precipitating factors. Positioning consideration is essential; patients’ bodies are often fused in a rigid position. All pressure points must be padded and the neck supported. If a patient’s cervical spine is fused in flexion, a steep Trendelenburg positioning is often needed for adequate dental exposure. Positioning considerations for Trendelenburg include padding the patients’ shoulders and securing the patients to the bed to ensure that their bodies do not shift on the table.

The authors of the 2011 “The Medical Management of Fibrodysplasia Ossificans Progressiva: Current Treatment Considerations”^a recommend the administration of perioperative corticosteroids to prevent and mitigate flare-ups. Corticosteroids were not administered in 11 cases. Long-term follow-up of the range of motion of the jaw was not

^aThe Medical Management of Fibrodysplasia Ossificans Progressiva: Current Treatment Considerations. Available at: <https://www.ifopa.org/living-with-fop-menu/treatment-guidelines.html>. Accessed March 4, 2013

obtained, but several patients who did not receive prophylactic perioperative corticosteroids reported postoperative jaw stiffness to one of the authors (FSK). Their jaw stiffness resolved after a 4-day postoperative course of corticosteroids for treatment of the flare-up. A 4-day perioperative corticosteroid course should be administered according to the current guidelines and begin before the start of the procedure.

In summary, a multidisciplinary approach to the perioperative management of patients with FOP should be the standard of care. Patients should be pretreated with corticosteroids and carefully positioned for surgery. GA can be safely administered to FOP patients for dental procedures; an awake nasal fiberoptic intubation is suggested as the first choice for airway management. Most patients can be discharged to home on the same day as their dental procedure.

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