Simulation-Guided Tracheotomy in a Patient With Fibrodysplasia Ossificans Progressiva

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Fibrodysplasia ossificans progressiva (FOP) is a rare disorder that causes heterotopic bone formation leading to chest wall and spinal deformities. This case describes an 11-year-old female with FOP who presented in respiratory failure necessitating two emergent fiberoptic nasotracheal intubations. The patient had severe trismus, rotary flexion of the neck, and distortion of the airway. A three-dimensional printed model based off of a computed tomography reconstruction was created for an in situ simulation before the true procedure. The surgery and trach change were both uneventful. We propose that with careful preoperative planning, tracheotomy can be an appropriate option for FOP patients.

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INTRODUCTION

Fibrodysplasia ossificans progressiva (FOP) is an extraskeletal disorder that causes progressive ossification of striated muscles spontaneously and in response to trauma.1,2 This is an autosomal dominant condition, although it frequently occurs from a spontaneous mutation.3 FOP tends to initially affect the neck and trunk within the first decade and then later progresses to distorting the lower limbs.4 Neck and truncal deformities lead to severe kyphoscoliosis that can increase the risk of restrictive lung disease, thoracic insufficiency syndrome, and eventually cardiopulmonary failure.1 In these patients, airway management is challenging; many times it requires nasal fiberoptic intubation due to severe trismus.5 Direct laryngoscopy is either impossible due to limited mouth opening or contraindicated because any manipulation of the temporomandibular joint by direct laryngoscopy could cause further ankylosis.

One study describes a tracheotomy for a patient who had ossification of the submandibular gland causing airway compromise; however, most of the literature advocates against a tracheotomy given the progressive nature of ossification with any soft tissue trauma and the difficulty in approaching the neck secondary to the anatomic distortion.1,6

We describe a case report of a patient with FOP who underwent tracheotomy placement. Due to the patient’s difficult anatomy, the patient received a computed tomography (CT) scan from which a digital three-dimensional (3D) model was constructed and a custom Bivona (Smiths Medical, Minneapolis, MN) tracheotomy tube was manufactured based on the measured dimensions. An in situ simulation using a 3D printed version of this model was conducted to aid the surgical and anesthesia teams with preparation and planning for the actual case. Due to the rarity of this disease and the even more rare instance of performing a tracheotomy, this case report may serve as a reference to providers who are faced with a similar situation. Written parental consent was obtained prior to submission of this case for publication. This report qualified for international review board exemption.

CASE REPORT

An 11-year-old female with FOP and severe kyphoscoliosis presented in status asthmaticus with acute hypoxemia and hypercarbic respiratory failure secondary to parainfluenza and was failing all medical management, including continuous nebulized albuterol, methylprednisolone, intravenous magnesium, terbutaline infusion, and nasal bilevel positive airway pressure (BiPAP). The patient was intubated by the otolaryngology service in the operating room via a fiberoptic nasotracheal approach. The patient was extubated to BiPAP 3 weeks later. The patient remained extubated until experiencing an aspiration event that necessitated a second fiberoptic...
nasotracheal intubation 1 month later. Multidisciplinary discussions between the pediatric intensive care unit, the family, and the otolaryngology service regarding a tracheotomy included ethical considerations, risks, and benefits. A CT neck and chest were performed in order to better evaluate the patient’s anatomy (Fig. 1).

Given the patient’s difficult anatomy, the radiology team was contacted regarding the creation of a 3D reconstruction to help better understand the patient’s cervical anatomy. Volume- and surface-rendered digital 3D models were created from the CT data (Fig. 2). The 3D reconstruction was reviewed with our multidisciplinary team to assess whether a tracheotomy was possible. The discussion, along with the specific risks, benefits, and alternatives of the procedure were presented to the family. After deliberation weighing speed of construction and cost, a 3D model was printed using bonded powder (Fig. 2), and a polyurethane elastomer was used to fill the cavity (Fig. 3). The model allowed for visualization and palpation of the vessels, visceral structures, and trachea using different colored pigments in the printed model.

On the day of the procedure, a 90-minute, just-in-time simulation scenario was scheduled before the surgery to be held in the same operating room and with the same care team that would be caring for the patient during the patient’s procedure. The team performed a fiberoptic intubation on a low-fidelity mannequin with a head and neck about the same size as the patient and simulated possible airway complications, including a plan of action for airway fire, loss of pilot balloon function, accidental extubation, and bleeding into the airway. Then, the 3D model was situated in the operating room with drapes to replicate the true operative procedure (Fig. 4). Upon dissection through the mold, the trachea was identified slightly more lateral than previously estimated. The landmarks on the 3D model aided in knowing where to place the incision and which landmarks to identify to help guide the surgical team to the trachea. In this patient, the trachea was just cephalad and deep to the right clavicular head. A simulation debriefing immediately following the scenario identified additional equipment, such as deeper, angled retractors, that was needed in the operating room and reviewed contingency plans for the team.

The additional equipment was obtained; the room was turned over; the patient entered the room; and the actual procedure was then performed with the same care team. The trachea was identified just cephalad and deep to the right clavicular head, as previously encountered on the 3D model. The right sternocleidomastoid muscle was densely fibrotic and had to be divided in order to visualize the airway. Stay sutures were placed, and the stoma was matured. The adjustable, custom-length, Bivona FlexTend tracheostomy tube (Smiths Medical) was inserted, and placement was confirmed with flexible tracheoscopy. Direct laryngoscopy was attempted with a Miller blade.

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Fig. 1. Preoperative imaging. (A) 3D reconstruction from a CT scan of the chest. (B) 3D reconstruction from CT scan of trachea (shown in blue) and great vessels (shown in brown). (C) Plain film chest X-ray. Note the extreme neck flexion to the left with deviation of the airway to the right.3D = three dimensional; CT = computed tomography.
laryngoscope, and the uvula was the only pharyngeal structure able to be identified. Further attempts using other laryngoscopes such as a glidescope were not attempted due to the very poor exposure obtained by the Miller blade laryngoscope and hesitation to induce trauma to the temporomandibular joint. The patient was then transported back to the pediatric intensive care unit and remained sedated for 2 days postoperatively. The care team then debriefed the actual procedure and specifically discussed the relevance of the just-in-time simulation.

Fig. 2. Digital 3D model shown on left (A and C). On the right (B and D), the bonded powder 3D model created from the digital model shows vessels (printed with lumens filled for ease of identification: artery: red, vein: blue, trachea: blue, bones = white). Small pegs can be seen holding the bones together and attaching the model parts to the frame to ensure positional stability of the internal structures when pouring the material. 3D = three dimensional; CT = computed tomography.

Fig. 3. Polyurethane elastomer (5 Shore) mold placed into frame. The outer frame (the outside of the skin surface) was then removed after the elastomer had fully cured (~12 hours).

Fig. 4. Setup of three-dimensional model for in situ simulation.
The patient received intravenous steroids the day of surgery and on postoperative day 1, followed by 2 days of tapering. On postoperative day 10, a bedside tracheostomy tube change was performed with a flexible scope at the bedside. The change was uneventful, and the stoma was mature. The scope exam revealed good position with minimal granulation tissue at the distal end of the trach. Tobramycin and dexamethasone drops were started through the tracheostomy tube; 2 days later, the granulation tissue appeared to be improving and the medication was discontinued.

At the writing of the article, the patient has become ambulatory after months of being bedridden, requiring ventilatory support at night only, and has not experienced any trach-related complications. The participants of the simulation included the anesthesia team, the otolaryngology team, the surgical technician, and the nursing staff. Simulation facilitation and debriefing were conducted by an experienced simulation facilitator. Surveys were filled out both after the simulation scenario and after the surgery.

DISCUSSION

We describe an uneventful tracheotomy following comprehensive surgical and anesthesia just-in-time simulation in a patient with fibrodysplasia ossificans progressiva. There has only been one study in the literature reporting a tracheotomy in a patient with FOP. Many reviews discourage this due to the difficult anatomy and potential for traumatic ossification. Unpublished communication with multiple institutions who have cared for patients with FOP and tracheotomies have described the potential for routine trach care without unusual complications. This case illustrates the effectiveness of 3D modeling and just-in-time in situ simulation for preparing for and performing a difficult tracheotomy in patients for whom there are no other options for airway management. The most common cause of death in patients with FOP is related to thoracic insufficiency syndrome causing pneumonia and right-sided heart failure7; therefore, if these patients can acquire a safe and secure airway, such as a tracheotomy, and receive positive pressure ventilation, perhaps earlier intervention may reduce the severity of morbidity and increase the age of mortality.

The concept of 3D models for operative planning is a method that has been used across disciplines with success.8 We propose that 3D models help simulate difficult anatomy, and in situ simulation can provide better preparedness for difficult cases. Of the seven postsimulation/presurgical surveys, over 75% of the participants agreed or strongly agreed that the simulation would help improve patient safety, teamwork, and evaluation of roles and responsibilities—and also identify equipment-related issues. Of the nine postsimulation/postsurgical surveys, all team members agreed or strongly agreed that the simulation aided in those same categories (Fig. 5). The summary response from the nine postsurgical surveys indicated that the simulation was either “invaluable” or “essential” in a case like this. The two members who did not fill out the presurgical survey were involved in both the simulation and the surgery.

Major concerns have centered around the altered anatomy secondary to contractures and kyphoscoliosis and the hypersensitivity to trauma-induced inflammation and ossification in FOP. In order to address these concerns, 3D imaging, modeling, and in situ simulation can not only be helpful for team preparation but may be important in order to better guide the dissection to reduce the extent of unnecessary tissue disruption during the procedure. Reconstructed CT images identified major structures such as the great vessels, clavicle, and sternum in relationship to the trachea. Our institution created a 3D external frame model made of bonded powder and filled the frame with polyurethane elastomer (5 Shore). The postsurgical survey comments suggested that this material was too dense when compared to human soft tissue. There are characteristics of the model material that are needed to best replicate soft tissue. These include force displacement measurements to monitor cutting force and material displacement after being

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Fig. 5. Simulation survey results.
It has been described that a more ideal model would have nonlinear material incompressibility, which would be compliant to small strains but resistant to more significant distortion. These are more expensive, and linear elastic models tend to be used because they are simple and efficient to create. An algorithm to create a more suitable 3D model material has been described. These considerations may be included in planning for future 3D modeled in situ simulation surgical rehearsals.

The other criticism of the simulation from the surveys was not having responsive hemodynamics that corresponded to the simulated airway emergencies. Although perceived to be important by some of the participants in this case, previous studies have demonstrated vital signs as being of low benefit in simulation. Overall, the simulation was perceived to be very helpful in the preparation for the actual case.

Wound care is a significant consideration for this patient population. Steroids have been shown to be effective in halting the inflammatory response in patients with FOP. Mouse models have shown that corticosteroids and the retinoic acid receptor gamma agonist palovarotene work to compete against heterotopic ossification in combination and individually. In humans, steroids and bisphosphonates have shown promising results to diminish the acute inflammation in the early period of a trauma. Nonsteroidal anti-inflammatory drugs and mast cell inhibitors have also been utilized, but no significant results have been reported. Our patient received a short course of steroids in an attempt to balance wound healing and the dampening of the inflammatory response, as described above.

Another consideration for wound care includes the actual tracheostomy tube and trach ties. Our institution uses velcro trach ties. Upon our first trach change, it was noted that the flare of the lateral aspect of the trach flange had created a grade-1 pressure ulcer on the inferior aspect of the chin, which was incapable of separating from the neck and chest due to complete neck extension immobility from a contracture (Fig. 6A). A custom tracheostomy tube was ordered with the straight flange (Fig. 6B), which allowed for less pressure on the submental skin and resolution of the ulcer in 7 days. Padding underneath the ties and inferior and superior to the face plate were placed, which is included in routine tracheotomy care at our institution. Although there was only a minimal amount of granulation tissue at the distal end of the tracheostomy tube, it is prudent to be cognizant that inflammatory tissue can ossify in these patients, creating a higher obstructive risk. It was encouraging to see this resolve with standard topical antibiotic and steroid-containing drops.

We present this case as the only complete description of a tracheotomy placement in a patient with FOP. We propose that 3D modeling and in situ simulation are pertinent and useful tools to decrease airway risk and more effectively manage a tracheotomy in select patients with FOP. This appeared to be an important factor in the safe performance of a tracheotomy procedure, which had been previously thought to be impossible in this patient. Wound healing is a critical factor that needs to be addressed as part of long-term care planning. Long-term survival of FOP patients after tracheotomy is unknown given the rarity of tracheotomies in this patient population. The novel application of 3D modeling and in situ simulation to aid in the preparation and planning for this type of procedure may be a useful tool for the surgical care team.

**CONCLUSION**

Fibrodysplasia ossificans progressiva is a disease process that inherently causes multiple airway difficulties for surgeons and anesthesiologists, often making tracheotomy difficult or impossible. The patient described above was deemed an appropriate candidate for a tracheotomy after extensive presurgical planning took place. Preparation using in situ simulation with a 3D model and team training in a just-in-time setting for the surgical case was beneficial to the entire care team. Simulation, as described in this case report, may make a positive impact on the severity of morbidity and age of mortality for these patients; however, future studies are needed to better evaluate its role in the surgical management of patients with difficult airways and FOP.
BIBLIOGRAPHY


