

# Anesthetic Management of Tracheo-Innominate Fistula in a Patient with Cerebral Palsy: Case Report and Literature Review

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## Abstract

Tracheo-innominate fistula (TIF) is a rare complication of tracheostomy placement with mortality rates from 73% to 100%. Focusing on anesthetic management concerns, we report a case of TIF in a patient with cerebral palsy and review the current literature on diagnosis and management of TIF to direct our recommendations.

## INTRODUCTION

Tracheo-innominate fistula (TIF) is a rare, often fatal, complication of tracheostomy placement. Estimated mortality rates range from 73% to 100%.<sup>1,2</sup> Initial management goals include maintaining airway patency, control of hemorrhage, and adequate cardiovascular resuscitation. Immediate diagnosis is critical for survival.<sup>3</sup> Control of bleeding and interruption of the innominate artery is the definitive treatment. Focusing on anesthetic management concerns, we report a case of TIF in a patient with cerebral palsy, review the current literature on diagnosis and management of TIF, and make recommendations based on our findings.

## CASE REPORT

A 29 year old, 44 kg, 5'4" female with cerebral palsy presented to the emergency room at our tertiary care hospital with a one-day history of intermittent hemoptysis. One month prior to presentation, the patient had received an emergent tracheostomy for respiratory failure. At her one week otolaryngological follow-up visit, deviation of the size 6 Shiley tracheostomy tube was noted. The deviation was attributed to the patient's neck contractures and dystonia. The patient was scheduled for a two-week follow-up visit, but began having intermittent hemoptysis at home requiring ambulance transport to the emergency room prior to her appointment.

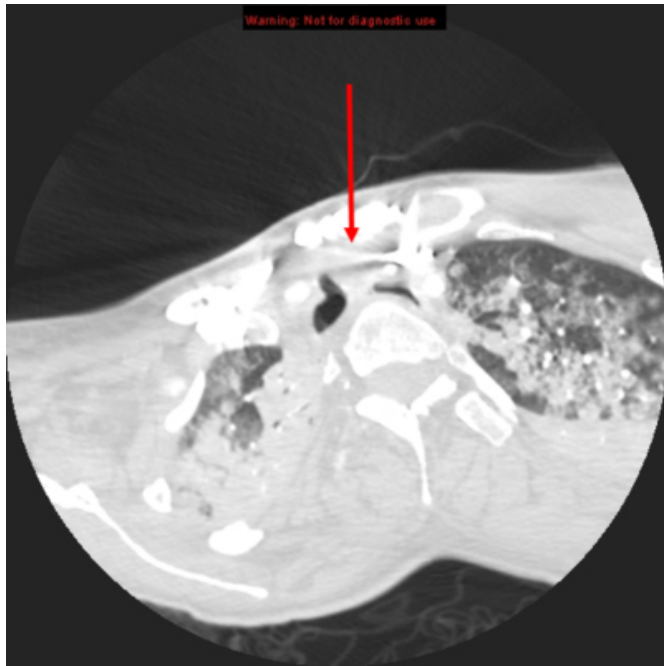
Upon arrival to the emergency room, no bleeding was seen at her tracheostomy site. The otolaryngology service was

consulted, and recommended that the patient be sent for CT angiography of the neck with IV contrast to localize the potential source of bleeding. Her hemoglobin at presentation was 13.6 gm/dL.

The CT angiogram showed irregular mucosa at the distal aspect of the tracheostomy catheter, representing focal ulceration or erosion. The tracheostomy tube was in place. No active extravasation was seen in or around the trachea. There was "diffuse upper lobe dependent airspace consolidation, likely related to the ongoing hemoptysis and aspiration," and "a non-specific, wedge-shaped area of hyperdensity within the peripheral right upper lobe" was noted in the radiologist's report. No definite vascular malformation or active extravasation was identified, and the cervical arterial vasculature was unremarkable (see Figure 1).

**Figure 1**

Figure 1. Distal Tracheostomy At Site of Ulceration



Following the CT angiogram, the patient began to have renewed hemoptysis in the emergency room. Despite an inconclusive CT scan, the otolaryngology team could not rule out a TIF based on this insensitive, non-specific study, and given her critical status and clinical presentation, she was scheduled for emergent neck exploration by the otolaryngology service to be performed with the cardiothoracic surgical team present. During her emergency room stay, the patient received 6 liters of fluids, 10 units of packed red blood cells, and had approximately 2 liters of blood suctioned from her tracheostomy site.

A femoral central venous line and right radial arterial line were inserted by the emergency room team, and the patient was orally intubated with a 6.0, cuffed endotracheal tube prior to transport to the operating room in an attempt to provide adequate airway access and to tamponade the bleeding with the inflated tube cuff. During her emergency room stay, the patient became severely hypotensive and required phenylephrine and a brief period of cardiopulmonary resuscitation. She was evaluated by our team in the emergency room, and given her critical status, we recommended that she be immediately transported to the operating room for surgical management.

Upon arrival to the operating room, the patient again became severely hypotensive with pulseless electrical activity requiring cardiopulmonary resuscitation. ABG in the

operating room revealed a pH of 6.98, pCO<sub>2</sub> of 48mmHg, pO<sub>2</sub> of 92mmHg, base excess of -19, lactate of 6.78 and hemoglobin of 7 gm/dL. Resuscitative doses of epinephrine (5mg), vasopressin (20 units), calcium (4 grams), along with fluid and blood transfusions resulted in systolic blood pressures of 56 to 80, but after 20 minutes she required cardiopulmonary resuscitation, which was continued for 25 minutes with continuous, pressurized blood loss from her endotracheal tube. Eight units of blood, one of FFP, and one 6-pack of platelets were transfused via a Belmont Rapid Infuser. The ventilator circuit was twice exchanged to prevent blood from backing up into the machine. We estimated approximately 1500 mL of blood loss from her endotracheal tube.

The otolaryngology service was unable to localize and control the bleeding at the level of the neck, so median sternotomy was performed by the cardiothoracic surgery team, and the perforated innominate artery was discovered near the distal tip of the tracheostomy tube. Upon isolation and compression of the innominate artery, the right radial arterial line tracing was lost, and we were forced to rely upon non-invasive blood pressure readings. Despite heroic measures including median sternotomy and cardiac massage by the cardiothoracic surgical fellow, adequate resuscitation and oxygenation with ventilation could not be achieved, and it was decided that further efforts or extracorporeal membranous oxygenation would be futile. The patient expired 7 hours after arrival to the hospital. No autopsy was performed.

## DISCUSSION

Given the emergent nature and high mortality rates of TIF, immediate diagnosis is the key step to survival for these patients. This complication occurs in less than 1% of all patients undergoing tracheostomy. Approximately 75% of cases will occur within 3–4 weeks of tracheostomy placement, with mortality rates approaching 100% even with surgical intervention.<sup>1</sup> Risk factors for the development of TIF include excessive movement of the tracheostomy, high-pressure (or overinflated) cuff, or a tube that has been placed too low.<sup>4</sup> TIF has also been reported as a complication of radiation.<sup>5</sup>

Upon review, we would not have recommended CT for diagnosis prior to surgical management of this patient, given that TIF is primarily a clinical diagnosis, with CT and other diagnostic exams being generally insensitive and non-specific.<sup>2-8</sup> In this particular case, the otolaryngology service

may have first ordered the CT angiogram and considered stenting the damaged vessel, once localized, given that the patient had no signs of active bleeding upon initial presentation to the emergency room. Diagnosis can usually be made at bedside without additional studies. In an analysis of 36 case reports, it was found that bronchoscopy was diagnostic in only 1 of 4 patients, and extravasation was seen in only 3 of 9 angiograms.<sup>2-8</sup> There have even been case reports of TIF's mimicking endotracheal masses.<sup>9</sup> The most common clinical presentations are bleeding around the tracheostomy tube or massive hemoptysis. Avoidance of prolonged or extreme hyperextension of the neck is helpful in preventing this fatal complication, and using lightweight tubing to avoid excessive downward pulling of the tube is also recommended.

Emergency digital or tube-cuff compression of the fistula can achieve hemostasis and allow for transport to the operating room for immediate surgical repair. The goal of surgery is to interrupt the innominate artery which, if successful, is associated with a low risk of re-bleeding (7%) and good long-term survival (64%). Maintenance of continuity of the innominate artery is contraindicated because of a high re-bleeding rate (60%) and poor long-term survival (10%).<sup>2</sup> Recently, several cases of successful endovascular repair of TIF have been reported in non-surgical patients.<sup>10-15</sup> Nonsurgical management in the pediatric population has been attempted as well, although surgical treatment is still the definitive treatment.<sup>16</sup> No evidence of significant neurologic or vascular compromise has been shown in association with interruption of the innominate artery, but there have been case reports of severe motor and intellectual disability subsequent to hemorrhage from the TIF itself.<sup>2,17</sup>

As a temporizing measure to decrease hemorrhage, a finger should be inserted into the tracheostomy site and attempt to compress the innominate artery,<sup>2</sup> with subsequent placement of invasive monitors including central lines and arterial lines as soon as possible. If a radial arterial line is placed, preference should be made for a left radial arterial line so that accurate blood pressure monitoring can continue following compression of the innominate artery.

Preparations for massive transfusion should be initiated, and adequate blood pressure should be maintained with blood products, fluids, and vasopressors as needed. If hemoptysis is present, oxygenation and ventilation may be difficult as blood fills the alveoli. Consideration could be given to

having cardio-pulmonary bypass or extra-corporeal membranous oxygenation available in case control of the bleeding artery can not be obtained, although we have not seen any evidence supporting this approach.

Patients with neuromuscular disease leading to dystonic neck contractures may be at a higher risk of TIF, given the abnormal neck anatomy leading to abnormal position of the tracheostomy tube, and it might be prudent from a surgical standpoint to place a cuffed tracheostomy tube with the knowledge that these patients may develop this fatal complication in the future. A cuffed tracheostomy tube would potentially have increased our ability to tamponade the site. Without the presence of a cuffed tracheostomy tube, we would recommend intubation with a cuffed endotracheal tube to attempt to tamponade the bleeding, and digital compression if this measure was unsuccessful.

Based upon what we have learned from this case, we recommend avoiding delays in management to obtain insensitive, non-specific tests such as CT scans or bronchoscopy<sup>2-8</sup>, and instead urge immediate surgical exploration and control of the bleeding vessel based upon the clinical diagnosis of TIF. We recommend that the patient be taken to the operating room at the first sign of bleeding, before hemodynamic instability occurs. Lines should be immediately placed in preparation for emergent resuscitation and potential cardiovascular collapse. Femoral central line access is preferred over subclavian or internal jugular lines (given the proximity of such lines to major vessels in the neck and thorax that may be compromised), and a left rather than right radial arterial line is preferred (given the potential need for compression of the innominate with subsequent loss of invasive blood pressure monitoring).

Although cases of TIF have been previously reported, we have not found any directly addressing anesthetic management, nor any that implicate a relationship between TIF and distorted neck anatomy as can be found in patients with cerebral palsy and other neuromuscular diseases leading to contractures and dystonia. We hope that this report will aid future anesthesiologists in successful management of patients with TIF, and help prevent the occurrence of this fatal complication in patients with cerebral palsy.

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