

# Tracheoinnominate Artery Fistula Formation in a Child with Long-Term Tracheostomy Dependence

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## Abstract

We report a fatal tracheoinnominate artery fistula (TIF) in a 13-year-old female patient with long-term tracheostomy tube dependence due to chronic respiratory failure. Thirteen years after placement of her tracheostomy tube, the patient experienced two separate episodes of sentinel bleeding prior to a fatal hemorrhagic event. Diagnostic evaluation after the sentinel events was mostly nonconclusive. This case highlights the risk of TIF in pediatric age group, even years after initial tracheostomy tube placement, and the need for a high index of suspicion for TIF when children present with unexplained tracheal bleeding.

## Keywords

- ▶ tracheoinnominate artery fistula
- ▶ tracheostomy
- ▶ sentinel bleed

## Introduction

Tracheoinnominate artery fistula (TIF) is an unusual, life-threatening complication of both open surgical tracheostomy and percutaneous dilatational tracheostomy, and its incidence varies from 0.3 to 4.5%.<sup>1,2</sup> The peak incidence of TIF is within the first 3 weeks of tracheostomy tube placement, with most cases reported within the first 18 months.<sup>3</sup> While the majority of cases of TIF have been reported in adult patients, a few cases of TIF in pediatric patients have been noted in recent years.<sup>4,5</sup> However, a recent study looking at complications related to tracheostomy in children did not find TIF and major bleeding to be an important complication of tracheostomy.<sup>6</sup> The increase in the number of infants and children requiring long-term tracheostomy support for the management of chronic respiratory failure in the United States raises the possibility that the incidence of TIF may increase.<sup>7</sup> We report a case of a pediatric patient, with a long-standing tracheostomy tube, who developed a TIF and associated hemorrhagic sequelae.

The institutional review board (IRB) determined our project does not meet the definition of human subject research under the purview of the IRB according to federal regulations.

## Case Report

The patient was a 13-year-old female patient with arthrogryposis multiplex congenita, static encephalopathy, spastic quadriplegia, profound intellectual disability of unknown etiology, complex myoclonic seizures, disuse osteoporosis with multiple prior bone fractures, gastrostomy tube dependence, and chronic respiratory failure with tracheostomy and mechanical ventilation dependency. The patient's tracheostomy was placed during early infancy and no major complications related to tracheostomy were noted except for occasional episodes of bacterial tracheitis. The patient had never experienced prior episodes of bleeding from the tracheostomy.

On the day of her presentation, the patient's home care nurse reported bright red blood from her tracheostomy and mouth after tracheal suctioning, which was atypical.

After emergency department evaluation, the patient immediately underwent laryngoscopy, bronchoscopy, and bronchoalveolar lavage by a combined team of pediatric otolaryngologists and pulmonologists. The procedures revealed bloody crust in the tracheostomy tube and distal trachea but no active source of bleeding. With these findings, the patient was diagnosed with a pulmonary hemorrhage

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and suspected pneumonia and was admitted to the pediatric intensive care unit (PICU) for treatment with intravenous steroids and antibiotics.

Further diagnostic evaluation was undertaken within 48 hours of admission, including repeat bedside bronchoscopy, computerized tomographic angiography (CTA) of the chest, and echocardiography. The bronchoscopy revealed areas of hypertrophic tissue on the posterior wall of the trachea and mild irritation of the tracheal mucosa, normal appearing bronchi, and no evidence of bleeding in the tracheobronchial tree. Echocardiography was unremarkable. Chest CTA did not show evidence of pulmonary arteriovenous malformations but did demonstrate the innominate artery to be in close proximity to the anterior surface of the trachea near the tip of the tracheostomy tube (▶**Fig. 1**). The patient's hemoglobin level was 7.5 g/dL, decreased from a value of 12.5 g/dL a few weeks prior to admission, but as she was otherwise asymptomatic and stable for the next several days, she was discharged home.

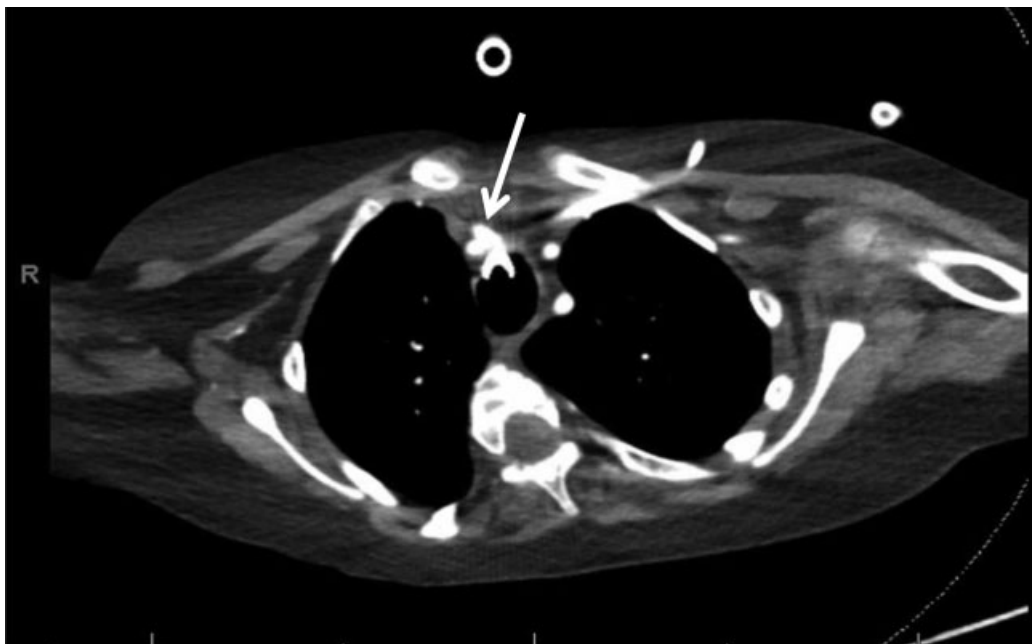
Three days following the discharge, the patient presented to the emergency department with another episode of tracheal bleeding. Therapy with intravenous steroids and antibiotics were resumed. Her tracheostomy tube was downsized from a 5.5-mm internal diameter and 46-mm length tube to a 5.0-mm internal diameter and 44-mm length tube because of the tracheal mucosal irritation noted during the prior bronchoscopy. Aggressive pulmonary hygiene therapy and tracheal suctioning were withheld. A brief episode of tracheal bleeding during a tracheostomy tube change led to a second airway bronchoscopy. It revealed mucous mixed with blood in the trachea and distal bronchi with no apparent bleeding source. The patient was readmitted to the PICU for further management. Her hemoglobin at that time had improved to 9 g/dL. Coagulation parameters and platelet count were normal.

Interventional radiology consultation was obtained, but no catheter-directed therapies were recommended based upon the results of the prior CTA. After 48 hours of stability, the patient was discharged to home.

Four days following the second discharge to home, the patient presented to an outside hospital following a sudden onset of large volume bleeding from the tracheostomy. Total blood loss by the time of PICU admission following transport was estimated to be 300 to 400 mL. Over the next hour, massive hemorrhage from the trachea continued resulting in hemorrhagic shock. Multiple blood products were administered. Pediatric cardiothoracic and general surgeons were consulted. Interventions included manipulation of the tracheostomy tube, inflation of the tracheostomy tube cuff to compress the area by repositioning of the tracheal tube, and direct arterial compression using a finger. Despite these interventions, she continued to bleed massively and progressed to cardiac arrest. She underwent emergent sternotomy with chest exploration, ligation of the innominate artery, and closure of the tracheal stoma. Ultimately, the patient had recurrence of cardiac arrest and spontaneous circulation could not be re-established.

## Discussion

TIF is a rare but devastating complication of tracheostomy tube placement. While reports of TIF have primarily involved adults with recent tracheostomy tube placement, reports like ours involving pediatric patients with long-standing tracheostomy tubes are emerging.<sup>4,5</sup> The expanding population of infants and children with tracheostomy tubes increases the possibility for TIF development. Therefore, clinicians who care for children with tracheostomy tubes must be familiar with this potential complication.



**Fig. 1** The CT angiography showing relation of innominate artery to tracheostomy tube (arrow). Innominate artery dividing just anterior to trachea and lower end of tracheostomy tube.

The innominate artery is the most commonly involved vessel in tracheal fistula formation after tracheostomy placement, but the common carotid artery, the innominate vein, the inferior thyroid artery, the internal mammary artery, and the aortic arch may also be involved.<sup>8</sup> The formation of a TIF is multifactorial and involves destruction of the anterior wall of the trachea and the posterior wall of the innominate artery. Mechanical issues that lead to fistula formation include high tracheostomy tube cuff pressures, a malpositioned tracheostomy tube tip, a high innominate artery, and a low-lying tracheostomy tube. Tracheostomy cuff pressures above 20 mm of mercury may result in pressure necrosis of the mucosa of the anterior tracheal wall.<sup>9–11</sup> Steroid use, stoma infection, malnutrition, excessive head movement, and spasticity also can exacerbate injury to the tracheal wall lying against the tip of the tracheostomy tube.<sup>12,13</sup> Our patient did not have a cuffed tracheostomy tube. However, she had other risk factors including spasticity and arching of the neck that may have led to long-term repetitive injury to the anterior wall of the trachea, multiple episodes of bacterial tracheitis and pneumonia, and multiple courses of steroid treatment.

Approximately 50% of cases of TIF have minor, bright-red preliminary (sentinel) bleeding before massive delayed hemorrhage. Sentinel bleeding is self-limited but recurrent in nature and is aggravated by coughing or aspiration. Ten mL or more of blood from an established tracheostomy should be considered diagnostic of a TIF and treated accordingly until proven otherwise.<sup>8</sup> Often, the sentinel bleed will spontaneously stop. Easy controllability of bleeding does not rule out the diagnosis of TIF. The mortality rate approaches 100% without urgent surgery as opposed to 20% with early recognition and proper management.<sup>14,15</sup>

If TIF is a possibility, bronchoscopy should be considered carefully as it can destabilize a clot that is preventing massive hemorrhage. Although bronchoscopy is unlikely to identify the fistula opening per se, it may exclude other pathology such as irritation, infection, erosion, or ulceration of tracheal mucosa. In addition, rigid bronchoscopy can clear the tracheobronchial tree of aspirated blood.<sup>8,16,17</sup> If bronchoscopy confirms that the main bronchi are free of blood, then immediate intervention is not required. However, further investigation to confirm other causes of bleeding, such as coagulopathy and bronchopneumonia, is vital. Other causes of hemorrhage must be excluded before a sentinel bleed associated with TIF can be confirmed.<sup>18</sup>

The small size of the fistula presents a major diagnostic challenge when using CTA for diagnosis. CTA may identify tracheal compression by an artery, intimate contact of an artery with the trachea, spilling of intravenous contrast into the airway, arterial erosion, and mediastinal infiltration, but actual visualization of a fistula with CTA has not been very successful. Thus, in the absence of active bleeding when sentinel bleeding had stopped, a CTA is not confirmative.<sup>19,20</sup> In the correct clinical setting where other causes of bleeding have been excluded, CTA evidence of close contact between an artery and the trachea at the site of the lower end of the tracheostomy tube may be sufficient evidence to diagnose a TIF such that, even with small sentinel bleeding, stomal exploration should follow.

If massive hemorrhage occurs, the goal is temporary hemostasis to allow transfer to the operating room. Over-inflating the tracheostomy tube cuff with the tracheostomy tube in a partially withdrawn position to directly occlude the bleeding vessel has been successful in 85% of cases. An oral endotracheal tube should be passed immediately with its cuff distal to the tracheal stoma to prevent blood from entering the airways. If bleeding is not controlled, the tracheostomy tube should be completely removed after oral endotracheal tube placement. In addition, digital pressure with the index finger should be applied on the innominate artery against the posterior surface of the sternum after dissection in the pretracheal fascia (Utley's maneuver). This maneuver is successful 90% of the time, and should be continued during transfer to the operating room, where surgical ligation of the innominate artery and fistula is the life-saving procedure.<sup>8,21</sup>

## Conclusion

TIF is a devastating and potentially fatal complication of tracheostomy tube placement. It may occur in long-term tracheostomy-dependent children, as in our case, but most cases described in the literature are reported following recent tracheostomy placement. There should be a high index of suspicion after any episode of bleeding from a tracheostomy in excess of 10 mL, as this may represent a sentinel bleed. Bronchoscopy and CTA might not confirm the diagnosis, but any close contact between any major artery or vein and the anterior tracheal wall on CTA in the appropriate clinical settings may be adequate to warrant urgent surgical intervention.

### Disclosure

None of the authors listed above had any real or perceived financial interest in relation to this case report.

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### Conflict of Interest

None declared.

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