

Congenital Tracheocutaneous Fistulas

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Objective: To outline two cases of congenital tracheocutaneous fistula and discuss the potential pathogenesis of this previously unreported developmental abnormality. **Methods:** Two cases of tracheocutaneous fistula evaluated at John Peter Smith Hospital (Fort Worth, TX) from May to October 2001 were reviewed. The surgical treatment of one infant is described. **Results:** Two infants were evaluated with a congenital fistula extending from the suprasternal region of the neck dorsally to the trachea in the midline. The infants were otherwise developmentally normal with unremarkable prenatal histories. Primary surgical closure of a fistula was accomplished without complication. **Conclusions:** Congenital tracheocutaneous fistula appears to be an isolated developmental abnormality not associated with the same degree of morbidity as acquired tracheocutaneous fistula. The development of a congenital tracheocutaneous fistula may be the result of abnormal epidermal migration secondary to a localized midline mesodermal defect. Congenital tracheocutaneous fistula may be successfully treated with primary closure. Observation and close follow-up of asymptomatic fistulas may be reasonable. **Key Words:** Congenital, fistula, tracheocutaneous, neonatal.

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INTRODUCTION

Persistent tracheocutaneous fistula (TCF) is a well-recognized potential complication following removal of a tracheotomy. It is thought to be secondary to epithelialization of the tracheocutaneous tract, preventing healing and closure of the fistula. Generally, surgical intervention is recommended for persistent fistulas, because of potential morbidity including skin or pulmonary infections, impaired ventilation, inability to tolerate submersion in water, and cosmetic deformity.^{1,2} In the present report, we present two cases of congenital TCF. We discuss the potential pathogenesis of this previously undescribed developmental abnormality.

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CASE REPORTS

Case 1

A 3.0-kg black male infant was born at 38 weeks' gestation by repeat cesarean section after an uncomplicated pregnancy. The infant initially displayed apnea complicated by the onset of bradycardia. Following positive-pressure mask ventilation of 15 seconds' duration, the patient achieved spontaneous respiratory effort with subsequent resolution of his cyanosis. The infant had no further respiratory or bradycardic event. Otolaryngologic examination was normal other than a 3-mm epithelialized fistula located in the midline suprasternal region (Fig. 1). The fistula tract was found to course dorsally in the midline when probed with an 18-gauge angiocatheter. Computed tomography (CT) scan revealed a 3-mm tract of air coursing from the suprasternal opening in the midline to the trachea with a collapsed intervening segment consistent with a TCF (Fig. 2). The infant was found to be otherwise healthy, and no other congenital abnormalities were present. The fistula was noted to be persistent at monthly follow-up examinations by the otolaryngology service. The infant remained otherwise healthy and was above average in weight and length at each visit. At 3 months of age the decision was made to proceed with repair of the TCF because there was noted to be persistent drainage from the tract. A lacrimal probe was easily passed through the fistula into the trachea to a depth of 14 mm. A skin incision was made circumferentially around the tract. Subcutaneous skin flaps were raised 1.5 cm superiorly and inferiorly. The fistula was dissected to the junction of the tract with the anterior tracheal wall (Fig. 3). The fistula was clamped and cut at its intercartilaginous entrance to the trachea in the midline. A 4-0 silk tie was used to ligate the dorsal base of the tract. Cervical fat was rotated to cover the oversewn tracheal defect and secured with a 4-0 Vicryl suture. The deep dermal layer was closed with 5-0 Vicryl, and the skin was sutured with 5-0 fast-absorbing gut. The infant did well postoperatively and was monitored overnight on the pediatrics floor. Histological evaluation of the fistula revealed that the majority of the fistula was lined with well-differentiated nonkeratinizing squamous cells. Along the dorsal fistula there was a clear transition to a lining composed of simple cuboidal cells (Fig. 4). He has been followed for 5 months postoperatively without any observed complications.

Case 2

A 3.6-kg Hispanic male infant was vaginally delivered at a gestational age of 39 weeks after an uncomplicated prenatal course. The infant was found to have a 2-mm sinus opening in the midline suprasternal region. The patient had no respiratory difficulties and was found to have no other abnormalities on examination. Probing with an angiocatheter revealed a tract coursing in the midline into the trachea. Computed tomography scan demonstrated the tract to be collapsed in the midline without air (Fig. 5). The abnormality has persisted at regular follow-up examina-

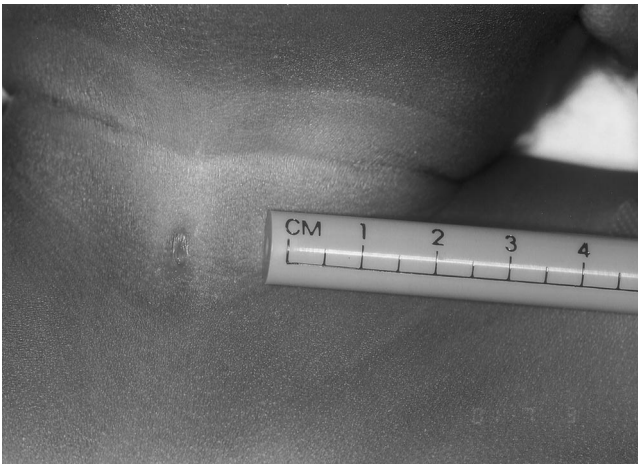


Fig. 1. Three-millimeter mm cutaneous opening of tracheocutaneous fistula.

tions with the otolaryngology service. The infant's family has deferred surgical treatment. The fistula has persisted at 4 months of age without complication. The patient has remained within normal limits for height, weight, and head circumference and has not been found to have any other physical abnormalities.

DISCUSSION

Congenital TCF has not been previously described. Similar to acquired TCF, congenital TCF appears to be resistant to spontaneous healing secondary to epithelialization of the tract. Both of our patients had persistence of the fistula for more than 3 months after birth. However, these two cases of TCF did not exhibit the same degree of morbidity as seen with acquired TCF. Neither infant had impaired ventilation or local skin or soft tissue infections. One infant did have mild drainage from the fistula. Both

children have been essentially asymptomatic except for parental concern regarding the cosmetic deformity. The diameter of the congenital fistulas is likely to be of inadequate size to allow significant alteration of ventilation or significant amounts of abnormal drainage of tracheal secretions. The walls of both congenital tracheocutaneous fistulas were collapsed on examination and required probing to reveal the dorsal extent of the fistula. Given the minimal pressure gradient between the skin surface and the tracheal lumen, only a small amount of force is required to overcome the high resistance of a narrow TCF to allow ventilation of the fistula. Following positive-pressure ventilation in the first patient, air was visualized in the fistula on CT scan. It is possible that increased tracheal air pressure facilitated the entrance of air into the collapsed fistula.

The pathogenesis of congenital TCF is not clearly evident. The laryngotracheal diverticulum develops as an endoderm-lined outgrowth from the wall of the foregut by the fourth week of embryogenesis. The endodermal lining of the laryngotracheal tube develops into the epithelium of the trachea and lungs. Splanchnic mesodermal tissue gives rise to the tracheal cartilage and surrounding connective tissue. The mesenchyme also determines the distal growth and development of the endodermal tissue. Recombinant tissue studies have shown that tracheal mesenchyme inhibits branching of tracheal endoderm, whereas bronchial mesenchyme promotes branching.³ In fact, fibroblast⁴ and epidermal growth factor⁵ homologues have been demonstrated to influence migration of tracheal cells in the *Drosophila* model.

A TCF could potentially result from abnormal ventral migration of endodermal tissue or from dorsal migration of ectodermal epidermis. A traumatic etiologic factor for congenital TCF is unlikely, given the uneventful prenatal



Fig. 2. Intraoperative view of patient in Figure 1, demonstrating lacrimal probe passing through tracheocutaneous fistula into tracheal lumen.

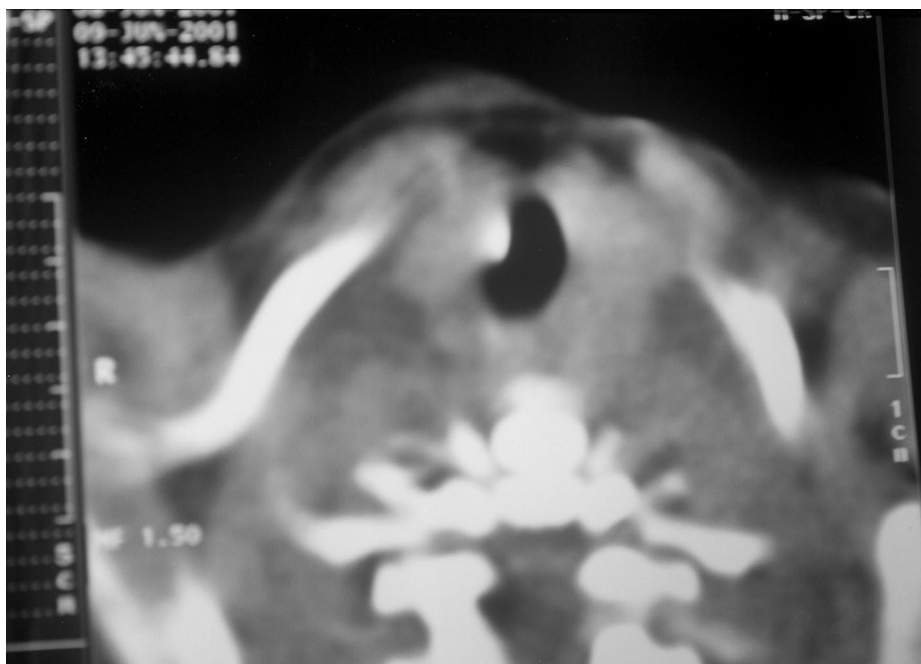


Fig. 3. Axial computed tomography scan of patient in Figure 1, demonstrating air pocket anterior to trachea with collapse of proximal portion of fistula tract.

course of the two infants. Neither mother underwent any invasive procedures such as amniocentesis. Because the fistula was lined by both squamous cells and simple cuboidal cells, it probably results from abnormal migration of endodermal and ectodermal tissue. Given that splanchnic mesodermal tissue normally intervenes between the epidermis and tracheal epithelium, a localized mesodermal defect may facilitate the dorsal migration of epidermis to the trachea. The proposed mesenchymal defect is isolated to the midline intercartilaginous connective tissue along the tract of the fistula. Neither patient was found to have abnormalities of tracheal cartilage on CT imaging or clinical examination. It is unlikely that the TCF developed from abnormal development of endodermal tissue because neither infant was found to have an aberrant ventral course of the tracheal lumen.

Other etiologic factors are possible as well. Tracheo-cutaneous fistula could conceivably represent a variant of the rare congenital midline cervical cleft disorder, breakdown of adhesions between the skin and trachea during development, or vascular ischemia leading to necrosis of an area of tissue between the skin and trachea during development. We think that these possibilities are less likely than the theory proposed above.

Congenital TCF appears to be an isolated, localized developmental abnormality. The patients were found to have no other congenital anomalies and have maintained normal postnatal growth and development.

There have been multiple methods used to describe the surgical treatment of TCF. Most techniques involve initial resection of the epithelialized tract. The wound can then be managed with dressing changes to promote healing by secondary intention^{1,2,6} or by primary closure. Several techniques have been used to augment closure of the fistula, including mobilization and closure of the strap muscles over the trachea,^{1,7} using the tract of the fistula

as a component of the closure,⁸ and performing a modified cutaneous Z-plasty over the repaired tracheal defect.⁹ Potential complications of primary closure including aerocele formation, subcutaneous emphysema, pneumomediastinum, and pneumothorax result from air leakage at the tracheal closure. Patients with large anterior tracheal wall defects¹ and with high tracheal air pressures secondary to upper airway obstruction¹⁰ are at a higher risk for morbidity with primary closure. However, a review of 98 pediatric patients with persistent TCF revealed no difference in postoperative complications between primary closure and healing by secondary intention.²

CONCLUSION

Congenital TCF is a rare, previously unreported, isolated developmental abnormality. The two cases described

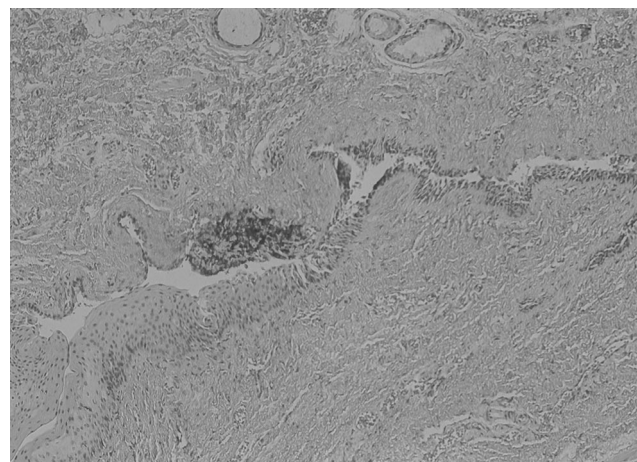


Fig. 4. Hematoxylin and eosin stain (original magnification $\times 50$) demonstrating transition zone between squamous-lined tract to cuboidal-lined tract.



Fig. 5. Axial computed tomography scan of second infant, demonstrating small defect in anterior tracheal wall with collapse of the distal portion of the tracheo-cutaneous fistula.

in the present study demonstrate that congenital TCF does not appear to be associated with the same degree of morbidity present with persistent TCF after decannulation. The pathogenesis of congenital TCF is not known but is hypothesized to be secondary to dorsal migration of epidermal tissue through a mesodermal defect in the midline of the lower neck. Resection of a congenital TCF with primary repair of the defect with a layered closure achieved an excellent result without morbidity.

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