The cause of the absent aortic valve remains unknown. Proposed causative factors include a deficiency of primitive valvular tissue, abnormal ventriculoarterial septation, and early destruction of the valvular leaflets. Because all reported patients are male, X-linked recessive inheritance or other X-linked factors have also been suggested.

The pathophysiology of this condition is complicated. Because the leaflets of the aortic valve are absent, free aortic valve insufficiency attributes to a persistent severely increased LVEDP, which promotes decreased coronary arterial perfusion pressure. Increased myocardial oxygen consumption caused by left ventricular hypertrophy, in combination with compromised coronary perfusion, produces a risk for ischemic heart disease.

As for echocardiographic findings, Bierman and coworkers[4] noted pandiastolic regurgitation in the aorta during the fetal stage, as demonstrated with Doppler echocardiography, and observed only an immobile unrestrictive ridge on the proximal endocardial surface of this valveless aorta immediately after birth. Because of the rapid hemodynamic deterioration after delivery, a prenatal diagnosis would facilitate neonatal critical care.

In our case the baby survived during the first palliative stage[5] because of the markedly diminished compliance of the left ventricle, which made it possible to maintain coronary perfusion. As the patient has grown, the left ventricle has developed with improved function. However, coronary perfusion might be sufficient because of the high LVEDP caused by mitral atresia and the absent aortic valve. In fact, cardiac catheterization showed aortic diastolic pressure and LVEDP to be almost the same. In addition, there was no pulmonary venous hypertension caused by mitral valve atresia or impaired oxygenation despite the increased left ventricular pressure.

We are observing the patient, paying special attention to the function of the left ventricle and the possibility of coronary steal syndrome.

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Congenital tracheoesophageal fistulas presenting in adults: Presentation of two cases and a synopsis of the literature

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More than 98% of tracheoesophageal fistulas (TEFs) are associated with atresia of the esophagus.[1] These appear in the neonatal period, and early surgical treatment provides a satisfactory result. In the group in which a fistula is not associated with atresia, termed H-type fistula, symptoms might be minimal, and presentation might be delayed until adulthood. We report a single surgeon's experience of 2 patients with this unusual condition. Bronchoesophageal fistulas are more commonly reported, with more than 140 such cases detailed in the literature.[2] We have reviewed and summarized the 14 reports in the English literature of congenital TEFs presenting in adults.

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Clinical Summary

PATIENT 1. A 45-year-old female nonsmoker had a recurrent productive cough. It was worse over the past 2 years and was particularly troublesome at night. Examination with a fiberoptic bronchoscope showed a punctum in the posterior wall of the trachea 3.5 cm from the carina (Figure 1). No fistula was identified on barium swallow or esophagoscopy. A cine contrast study done while the patient was in the prone position confirmed the fistula between the esophagus and the lower trachea. At right-sided thoracotomy, the fistula was identified, divided, and repaired. A flap of parietal pleura was used to augment the esophageal repair. The postoperative course was uneventful, and at 1 year's follow-up, she remained symptom free.

PATIENT 2. A 55-year-old man had a history of persistent cough since childhood. Throughout his adult life, he had recurrent bouts of bronchitis. He also noticed that he often coughed or choked after swallowing liquids. He had right-sided pneumonia just 3 weeks before admission. A chest x-ray film showed middle-lobe consolidation. Examination with a flexible bronchoscope showed a fistula

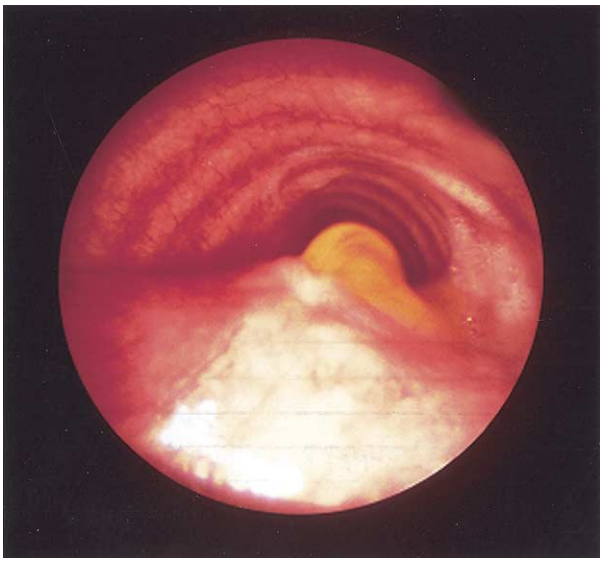

Figure 1. Bronchoscopic finding of a congenital TEF.

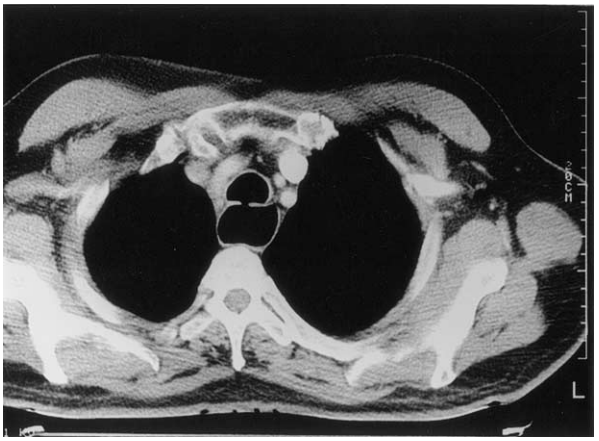

Figure 2. Computed tomogram of a congenital TEF.

in the posterior wall of the trachea at its midpoint. Barium swallow did not identify the fistula. Esophagoscopy showed the entry point to lie 20 cm from the incisors. This fistula was clearly seen on the subsequent computed tomographic scan (Figure 2), which was performed to assess parenchymal damage.

A left cervical incision along the inner border of the sternocleidomastoid muscle was used to access the fistula. Once identified, the fistula was divided, and both ends were repaired. A sternocleidomastoid muscle flap was fixed between the trachea and esophagus to reinforce the repair. Postoperatively, he did well, and at follow-up (6 months postoperatively), he remained symptom free.

Discussion

Congenital TEF in adults is infrequently described in the literature. We could find only 14 treated cases reported. It was first reported on a postmortem specimen by Negus[3] in 1929. It was only in 1954 that a case of thoracotomy and repair was reported.[4] Since then, patients have been successfully treated, and different approaches have been sporadically published.[1,2,5-8] The age of presentation ranges from 15 to 63 years, with an equal male/female ratio.

A chronic cough and recurrent respiratory infections are the common presenting symptoms. Bouts of coughing when swallowing liquids are pathognomonic for this condition.[2] A nocturnal cough and hemoptysis are other symptoms associated with this condition. Close questioning often leads to the history of symptoms being traced back to childhood. Delayed presentation might be related to gradual reduction in tone of the lower esophageal sphincter, with age leading to increased reflux and hence soilage of the trachea. Another plausible explanation is that of Bekoe and coworkers,[5] who suggested that initial mild symptoms are not properly investigated until complications appear.

A variety of investigations have been used in the diagnosis of TEFs. A chest x-ray film showing air distention of the esophagus must raise suspicion.[6] A thin barium swallow, particularly if done with the patient lying prone, often confirms the diagnosis. Bronchoscopy is useful at establishing the exact nature and type of fistula. Fistulas can be missed on esophagoscopy because they are in the upper third and on the anterior wall.[6] A computed tomographic scan is helpful at looking at the lung parenchyma to assess the extent of irreversible damage. It sometimes shows the fistula clearly (Figure 2).

There is no controversy about the necessity for surgical management of congenital TEF.[2] We used 2 different approaches to deal with these cases. Thoracotomy appears to be the most commonly used in the literature (10/14). It is the approach of choice when dealing with damaged lung, which might need resection. A cervical approach along the anterior border of the sternocleidomastoid muscle is more than adequate for most simple TEF repairs (3/14). Of the cases reviewed, the fistula could have been excised through a cervical incision in several of the cases. Because most H-type TEFs occur in the upper half of the trachea, a cervical incision provides an excellent exposure for repair. An upper midline limited sternotomy has been mentioned[7] but is not often required. A method to aid in identification of the fistula with a flexible guidewire across it at the time of the operation has been described,[8] but we did not find it necessary at the time of our dissection. After the dissection and division of the fistula, a muscle or pleural flap is recommended to prevent relapse.[1,2,7,8] We used it to protect the repair in both our cases.

We present this single surgeon's experience to highlight the different approaches available and the difficulty associated with diagnosis of this condition in this age group. Once diagnosis is confirmed, we recommend early surgical intervention because it is well tolerated, with complete resolution of symptoms. In case of a completely destroyed lung, resection is warranted.

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