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Editorial note by W. E. Berdon on the following paper by R. J. Hernandez and G. F. Tucker

This paper is an excellent one to draw one's attention to the easily missed diagnosis of tracheal stenosis, either intrinsic or extrinsic, due to compressive masses or vascular anomalies.

It stresses CT - in the review process, the subject of *cine CT* came up (now available in two Children's Hospitals in the United States) and undoubtedly MRI (with cine display) will be available, but not to everyone.

Every X-ray department has a fluoroscope, and a filter (of brass or tin-copper aluminum) can be made. The trachea, with this method, must be seen (with removal of bony obscuration by the filtration). An "empty segment" is due either to intrinsic or extrinsic disease, with stenosis versus collapse being the issue in infants.

It is the *establishment of the diagnosis, not the method used,* that is the important issue.

Congenital tracheal stenosis: role of CT and high kV films

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Abstract. Congenital tracheal stenosis is an intrinsic narrowing of the tracheal lumen due to complete cartilaginous rings. We evaluated the role of the chest radiograph, high kV films, CT and bronchoscopy in five patients with congenital tracheal stenosis. CT was superior to chest radiography and high kV films in the evaluation of the presence and extent of the stenosis. CT was superior to bronchoscopy in the evaluation of the distal extent of the stenosis in two patients. In addition, CT is useful in ruling out external compression of the trachea by a mass or associated vascular anomaly.

Congenital tracheal stenosis is defined as an intrinsic narrowing of the tracheal lumen due to complete cartilaginous rings. The tracheal cartilages are smaller than normal, firm, nonpliable and lack the normal structure of the posterior membranous trachea [1]. The walls of the trachea are rigid and nondistensible [2].

Several reports have described the use of computerized tomography (CT) in the evaluation of the trachea [3-9].

The purpose of this report is to compare the use of conventional radiographs of the chest, high kV films, CT and bronchoscopy in the evaluation of congenital tracheal stenosis. New surgical techniques for correction of the stenosis [10] require delineation of the extent of the involved stenotic segment for appropriate surgical correction.

Material and methods

We studied five children with congenital tracheal stenosis at Children's Memorial Hospital, Chicago. Their ages, sex distribution, and clinical findings are summarized in Table 1. The radiographic evaluation consisted of chest X-ray, high kV films of the airway and computed tomography of the neck and chest. One of the pat-

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Table 1. Summary of clinical, radiographic and surgical findings

	Age	Sex	Clinical findings	Chest X-ray	High KVP films	CT	Bronchoscopy	Surgery	Follow-up	Associated findings
1 RW	7M	M	2 episodes of severe respiratory distress at home	Not diagnostic	Narrowing 2 cm below the cords to 1 cm above carina	Narrowing 2 cm below the cords to 1 cm above carina. Caliber stenosis 4 mm	Tracheal narrowing beginning 1.5 cm below cords. Distal extent not determined. Caliber of trachea at site of stenosis 4 mm	Tracheoplasty extent of stenosis same as demonstrated radiographically	Asymptomatic 2 years after surgery	Tracheal right upper lobe bronchus
2 PM	2Y	M	Repeated episodes of respiratory distress	Localized narrowing of trachea above sternoclavicular joints	Localized narrowing of trachea above sternoclavicular joints	The caliber of trachea at stenotic site 8 mm. The length of stenosis 2 cm. Caliber of trachea beyond stenosis 11 mm	Narrowing extended 2–2.5 cm. Caliber of trachea at narrowing site 6 mm	Tracheoplasty. Findings identical to conventional radiographs and CT	Discharged symptom free	
3 RD	4M	F	Respiratory distress and possible aspiration	Poor visualization of trachea	Poor visualization of trachea	Narrowing extending from 1 cm above sternoclavicular joint to carina. Lgth stenosis 3.5 cm; caliber stenosis 2 mm	Length of stenosis 3.0 cm. The internal diameter of stenosis 3 mm	Length of stenosis 3.5 cm	Patient left hospital 3 weeks after surgery symptom free	Dimple above carina may be residue of previous fistula
4 AH	21M	F	Since 8 weeks prior to admission several episodes of croup. On the night prior to admission she had an episode of coughing, vomited and became febrile	Opaque left hemithorax thought to be secondary to foreign body. Trachea difficult to visualize	Not taken. Patient intubated making it difficult to obtain films	Narrow trachea (0.3 × 0.5 cm) extending 2 cm below cords to carina. Agenesis of left lung. Left bronchial stump	Tight stenosis (2 mm) beginning 2 cm below the level of the cords. Distal extent could not be determined	Stenosis starting 2 cm below cords all the way to the carina	Patient required tracheostomy and ventilatory support for a month after surgery. 4 months after surgery pt extubated; remains well	Agenesis of left lung. Left bronchial stump. No left pulmonary artery
5 GJ	10M	M	Recently noted to have noisy breathing	Suggestion of tracheal narrowing	Narrowing of trachea beginning 2 cm below the cords to 2 cm above carina	Narrow trachea (1 mm) in diameter extending 35 mm	Stenosis beginning at cricoid to carina. Caliber stenosis 3 mm	The stenosis extended from the ring below the cricoid to the carina	Patient had a short postoperative period and is doing well	

ients did not have high kV films because of endotracheal intubation. The high kV magnification films were obtained according to the technique described by Joseph [11]. CT examination was performed using the EMI 5005 in three patients and using a GE 9800 in two patients. The thickness of each section was 1 cm and sections were obtained at 1 cm intervals. The diameter of the trachea in the CT slices was measured using the electronic calipers available in the CT scanner. The measurements were done with the sections displayed in the mediastinal settings (low positive level and a moderately narrow window width). The presence of stenosis was established when the diameter of the trachea diminished in relation to the subglottic region.

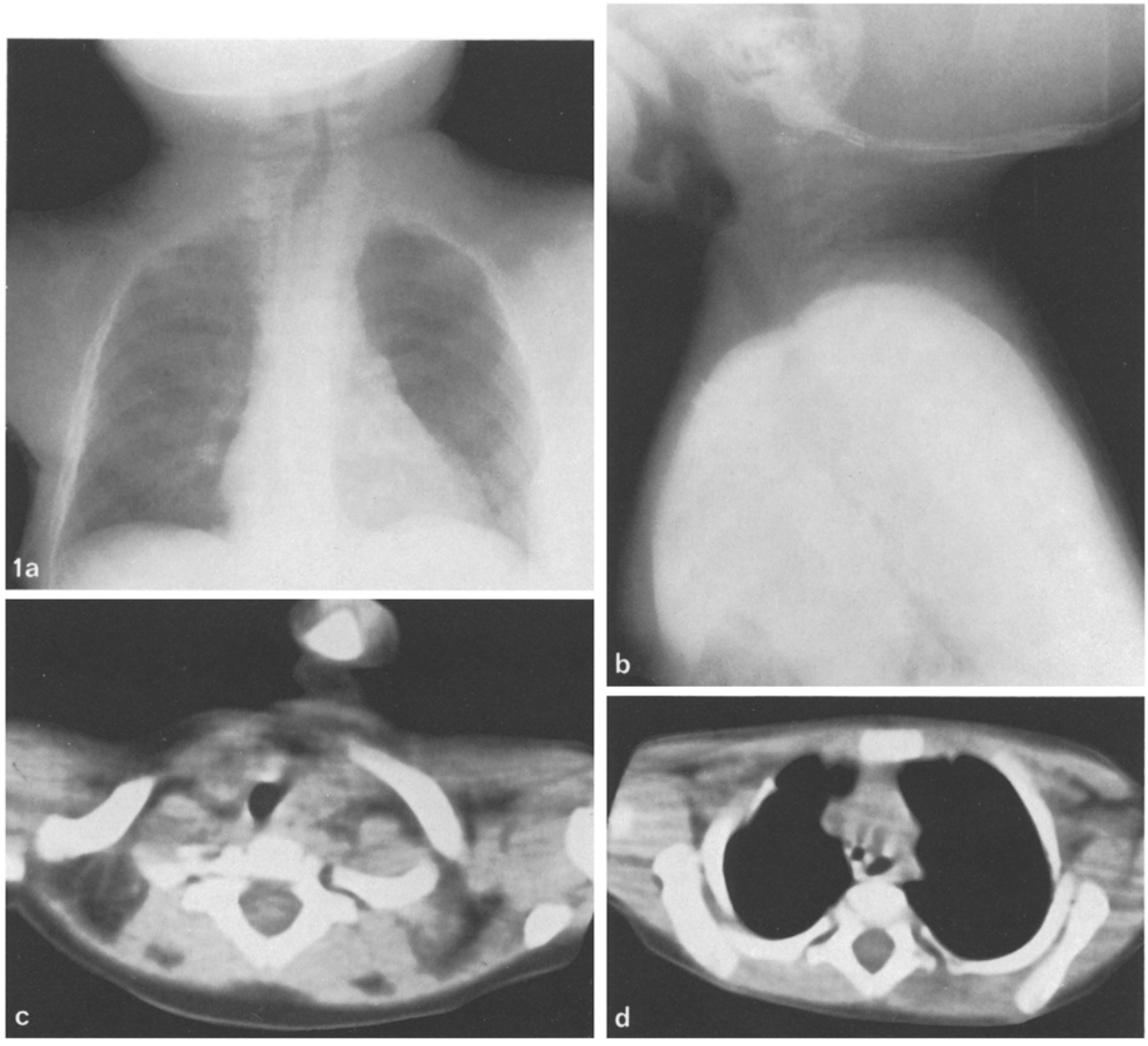


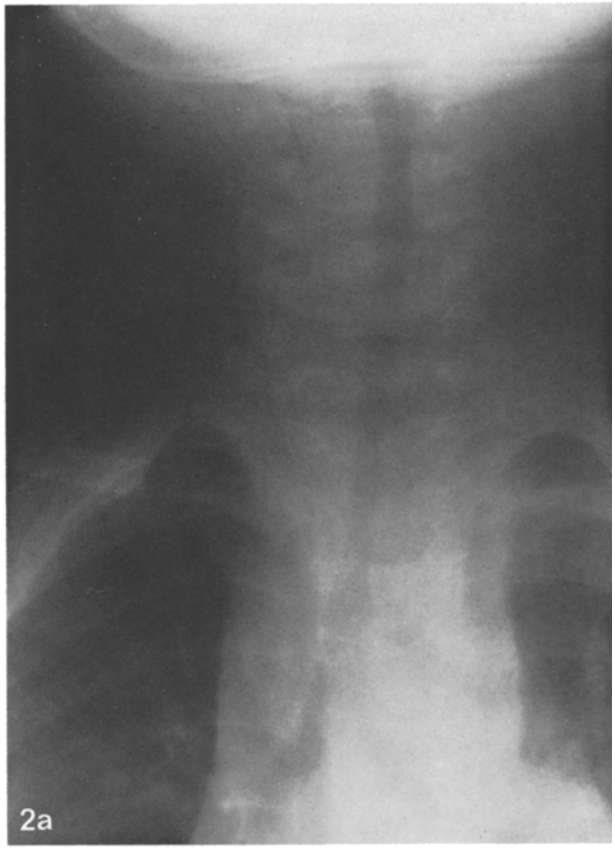
Fig. 1 a–d. RD Four-month-old female with respiratory distress. High kV film of the trachea in the frontal **(a)** and lateral **(b)** projections. The extrathoracic portion of the trachea is well visualized. Although there is a suggestion of narrowing in the intrathoracic portion of the trachea the visualization of the trachea is suboptimal despite several attempts. **c** CT of the trachea two centimeters above the sternoclavicular joint. The caliber of the trachea is normal. The cross section of the trachea measures 0.26 cm^2. The diameters of the trachea are 0.5 cm and 0.7 cm. **d** CT of the chest at the level of the aortic arch demonstrates the tracheal lumen concentrically reduced with a circular appearance. The cross section of the trachea at this level is 0.04 cm^2. The tracheal diameter is 0.2 cm. The air-filled esophagus is visualized posterior to the trachea

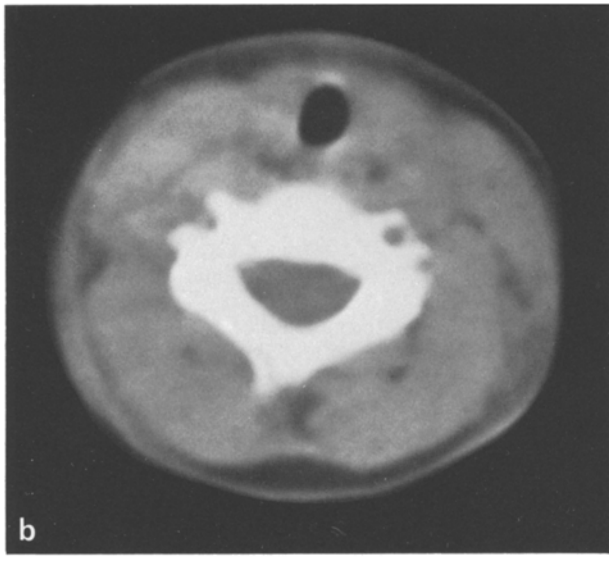
All patients had bronchoscopy and surgical confirmation of the presence and extent of stenosis.

Results

A summary of the clinical data, radiographic and surgical findings and follow-up is given in Table 1.

Of the four patients who had high kV films, the presence and extent of stenosis could not be determined in one (RD) (Fig. 1.), while they were diagnostic in the remaining three patients (RW, PM, GJ) (Fig. 2.). The chest radiograph was not diagnostic in one patient (AH) in whom high kV films were not obtained. CT was accurate in the assessment of the degree of stenosis and extent in all five cases. Bronchoscopy was unable to evaluate the length of the stenosis in two patients due to the marked narrowing of the trachea (AH, RW).





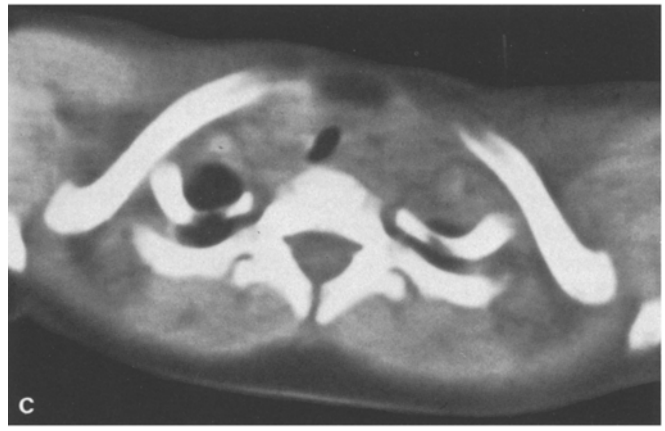


Fig. 2a–c. G. Y. Ten month old boy who recently was noted to have noisy breathing. **a** High kV films in the frontal projection. The trachea appears narrow from 2 cm below the vocal cords to 2 cm above the carina. **b** CT of the trachea in the subglottic region. The trachea is normal with a diameter 0.6 cm. The cross-sectional area measures 0.40 cm^2. **c** CT through the area of narrowing. The cross section of the trachea has an ovoid appearance. Although the greater diameter measures 0.5 cm, the cross-sectional area was reduced to 0.20 cm^2

Discussion

Congenital tracheal stenosis is defined as an intrinsic narrowing of the tracheal lumen; the walls of the trachea being rigid and nondistensible [2]. The tracheal cartilages are smaller than normal, firm and nonpliable, and lack the normal structure of the posterior membranous trachea [1]. In a series reported by Benjamin [2], 90% of the cases presented before the age of one year.

The clinical presentation is varied but often includes wheeze, respiratory distress and stridor. There are often associated anomalies of the respiratory tract and esophagus. Associated anomalies include bronchial stenosis, tracheal bronchus, hypoplasia or agenesis of the lung, pulmonary sling, and H-type tracheoesophageal fistula.

CT has been used in the evaluation of tracheal disorders such as tracheal stenosis [3, 4, 9]. Some of these reports deal primarily with tracheal stenosis secondary to previous intubation in the adult [3] and subglottic stenosis in the neonate [4]. The CT appearance of tracheal stenosis usually consists of a perfect circle (Fig. 1 d.) with a smaller diameter than the immediate subglottic portion of the trachea [9]. Although this circular appearance of the stenotic trachea is the most common, other shapes such as an oval configuration can also be observed (Fig. 2 c.). The shape of the trachea is not as useful as the diameter, since the trachea in children may be circular and often does not have the flattening of the posterior wall usually seen in adults [6].

CT and high kV films are useful in determining the extent of the stenotic segment with CT being superior to high kV films in our patients. This information is sometimes difficult to obtain by bronchoscopy due to inability to bypass the stenosis. An

alternative is to perform a tracheogram in a child with an already compromised airway but this is dangerous. Another approach is to evaluate the trachea by fluoroscopy but this usually results in a high radiation dose [4]. Intraoperative determination of the extent of the involved segment of trachea is difficult; thus the need of adequate preoperative evaluation.

External compression of the trachea by mediastinal masses (i. e. lymph nodes) can mimic the appearance of tracheal stenosis [8]. In addition tracheal stenosis can be associated with a pulmonary artery sling [12]. Contrast enhanced CT can diagnose these two entities [13, 14].

There are standards regarding the diameter [5] and the cross-sectional area [7] of the trachea in children. Since 90% of the cases reported by Benjamin [1] presented before one year of age, these standards are not useful since they are based on a very small sample in this age group.

In all our patients the immediate subglottic portion of the airway was not affected. This area, the conus elasticus, where tracheal cartilaginous rings are not present, is usually of normal caliber. The lack of involvement of the conus elasticus provides an internal standard with which to compare the stenotic trachea. It is important not to alter the window width or level while evaluating the caliber of the trachea since changes in the display will result in a difference of caliber. On the other hand, in subglottic stenosis [15] the narrowing is localized to this area and does not extend into the trachea.

One of the entities to be distinguished from tracheal stenosis is tracheomalacia. The anterior and posterior walls of the trachea come closer together due to the lack of stiffness with a reduction of tracheal lumen [16]. Fluoroscopy is helpful in evaluating these children since it will demonstrate changes in caliber of the trachea during different phases of respiration.

In summary, CT and high kV films of the trachea are useful in the evaluation of the presence and extent of congenital tracheal stenosis. These two imaging modalities can delineate the extent of the stenosis which sometimes is difficult to ascertain by bronchoscopy and avoid dangerous bronchography. In addition, CT can rule out narrowing of the trachea secondary to mediastinal masses or in association with a pulmonary sling.

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