

Tracheobronchomegaly

ZAKI AL-MALLAH¹ AND O. P. QUANTOCK

From the University of Mosul, Iraq, and Sully Hospital, Wales

Two cases of the rare condition of tracheobronchomegaly are reported. They occurred in people of completely different racial origin and residence. They showed the characteristic features of this condition—loud, rasping, prolonged, remarkably ineffective cough, abnormally wide trachea and major bronchi, laxity of the cartilaginous rings and membranous part of these airways demonstrable on straight chest radiographs and bronchoscopy and confirmed at bronchography. Evidence is submitted of congenital aetiology.

Tracheobronchomegaly is a rare congenital abnormality with marked widening of the trachea and major bronchi, in most cases associated with chronic recurrent respiratory tract infection. This rare syndrome was first described by Mounier-Kuhn in 1932, but was then given different names until Katz, LeVine, and Herman (1962), in an excellent review, clearly defined this entity and suggested the name tracheobronchomegaly. We have been unable to find a report of this syndrome in the British literature and are therefore presenting two cases, one occurring in an Iraqi living in his own country, the other in an Italian living in Britain.

CASE REPORTS

CASE 1 N. H., a 24-year-old male farmer from Jazira-Mosul, was admitted to Mosul Hospital, Iraq, in December 1964, complaining of severe productive cough with hoarseness and haemoptysis.

He gave a history of chronic productive cough for 10 years with sputum occasionally tinged with blood, worse during the winter months but clearing without treatment.

One month before admission he had a severe attack for two days with fever, chest pain, hoarseness, and a troublesome loud cough, productive of about 4 oz. (120 ml.) of mucopurulent sputum, sometimes blood-stained. He had a course of penicillin and streptomycin injections from his general practitioner and improved.

On admission the hoarseness persisted with prolonged bouts of a rasping, noisy cough, productive of sticky yellow sputum. The patient felt more comfortable in the prone position. He was a non-smoker. The family history was irrelevant. He was a well-developed,

apparently healthy young man with no finger clubbing. There were scattered wheezes and bilateral basal crepitations, mainly anteriorly. No abnormality was detected in any other system. The tuberculin test (100 T.U.) was negative and a chest radiograph showed small nodular opacities in the right lung; these subsequently cleared. Bronchoscopy showed reddening and oedema of the mucosa which was covered with thick, mucopurulent secretions. No intrinsic lesion was seen in the tracheobronchial passages, but an abnormal mobility of the posterior tracheal wall was noted.

A bronchogram (Figs 1 and 2) showed gross dilatation of the trachea and the main bronchi, not detected bronchoscopically. Irregularity of the bronchial calibre and mucous gland dilatation were also noted. The diameter of the trachea was 46 mm., of the right main bronchus 28 mm., and of the left main bronchus 32 mm.

The patient was treated with antibiotics and postural drainage, but when he improved he took his own discharge. No follow-up was possible.

CASE 2 M. G., a 44-year-old Italian, was admitted to Sully Hospital complaining that for seven years he had had exertional dyspnoea and difficulty in coughing up small plugs of sputum. He had also noticed a peculiar 'purring' quality to his breathing and he had to massage the trachea to expectorate. His voice had become hoarse. These symptoms had slowly been getting worse.

On examination he was obese and more comfortable in the prone position. His cough was protracted, very harsh, loud, rasping and noisy, waxing and waning for about 3 seconds instead of the usual sharp explosive cough noise. Indirect laryngoscopy was normal. All laboratory investigations were normal.

The only abnormality seen on the chest radiograph was a wide trachea. Tomographs of the trachea confirmed this (Figs 3 and 4). Bronchoscopy revealed a very unusual feature, that is, during expiration the posterior wall of the trachea moved forwards and

¹ Present address: The Welsh National School of Medicine, Tuberculosis and Chest Diseases Department, Sully Hospital, Penarth, Glamorgan

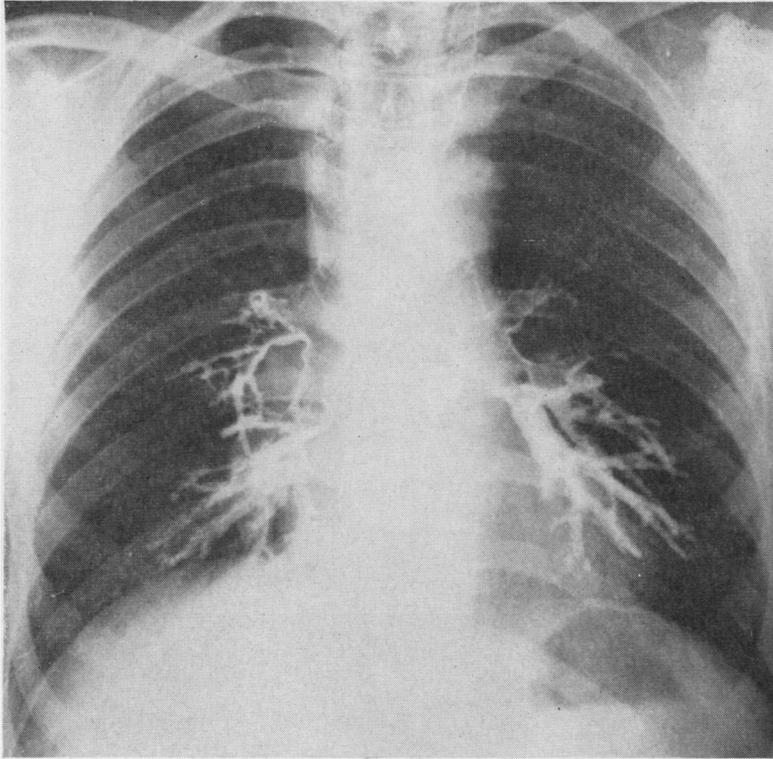


FIG. 1

FIGS 1 and 2. Case 1. Bronchogram, P.A. and right lateral views, showing the striking dilatation of the trachea and the main bronchi with irregular ectasia of the bronchial tree.

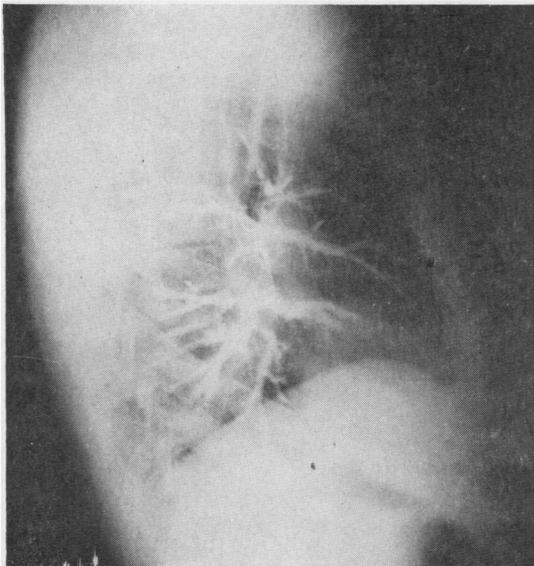


FIG. 2

almost totally occluded the lumen. During inspiration, however, the trachea ballooned out into an enormous cavernous space. A bronchogram (Fig. 5) showed the increased diameter and irregularity of the trachea and main bronchi. Tracheograms taken during inspiration and expiration (Figs 6 and 7) showed a marked variation in the transverse diameter of the trachea at the different phases of respiration.

The trachea measured 40 mm. at its greatest diameter, the right main bronchus 32 mm., and the left main bronchus 24 mm.

DISCUSSION

Both these patients presented with chronic respiratory symptoms. Their cough was extraordinary, being loud, rasping, long, and ineffective. Both had a very wide trachea. Bronchoscopically there was gross exaggeration of the normal calibre change on inspiration and expiration. Bronchograms confirmed the diagnosis.

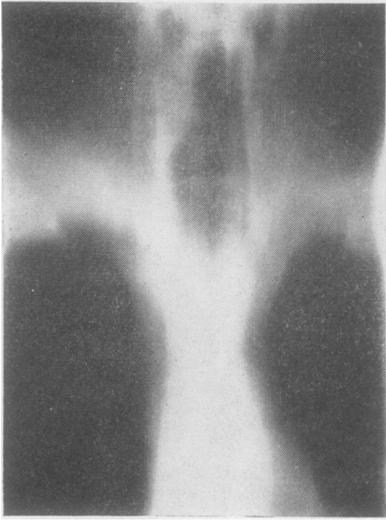


FIG. 3

FIGS 3 and 4. Case 2. A.P. and lateral tomograms showing the massive and irregular dilatation of the trachea.

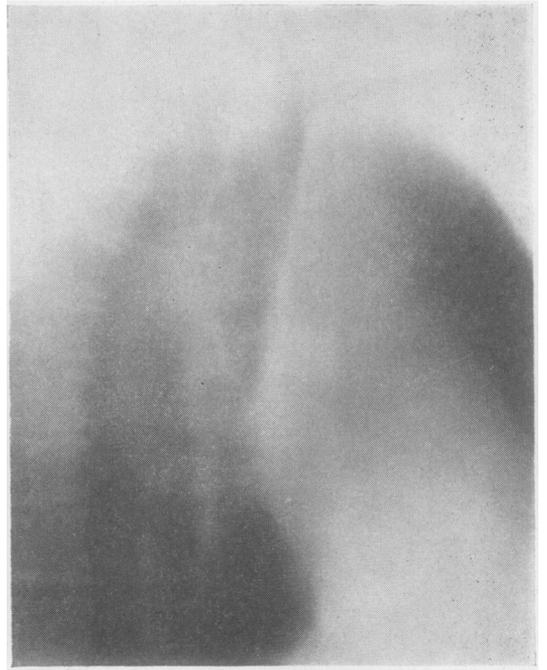


FIG. 4

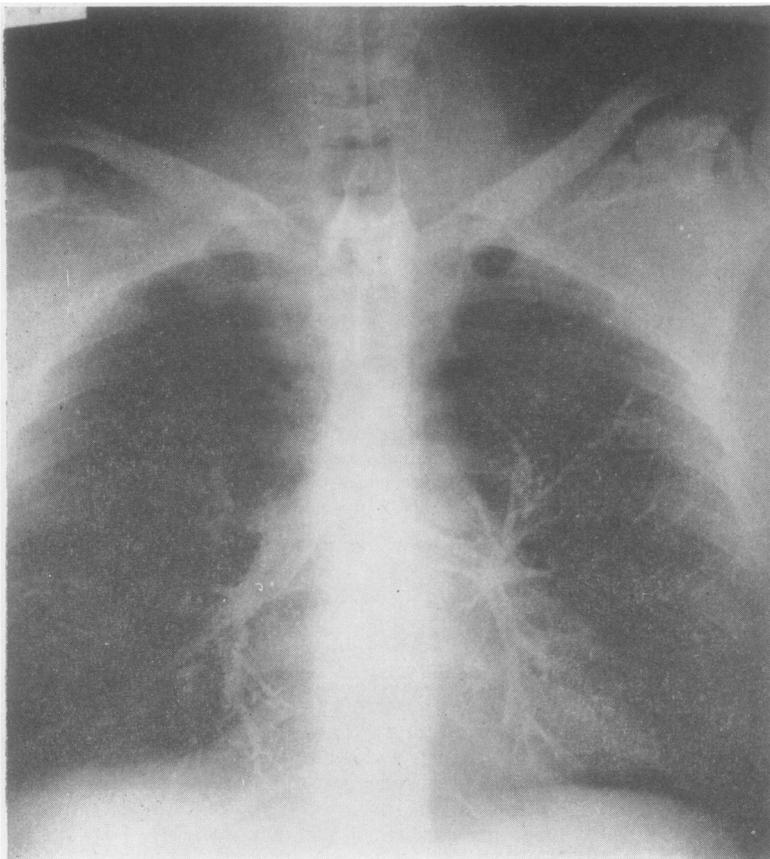
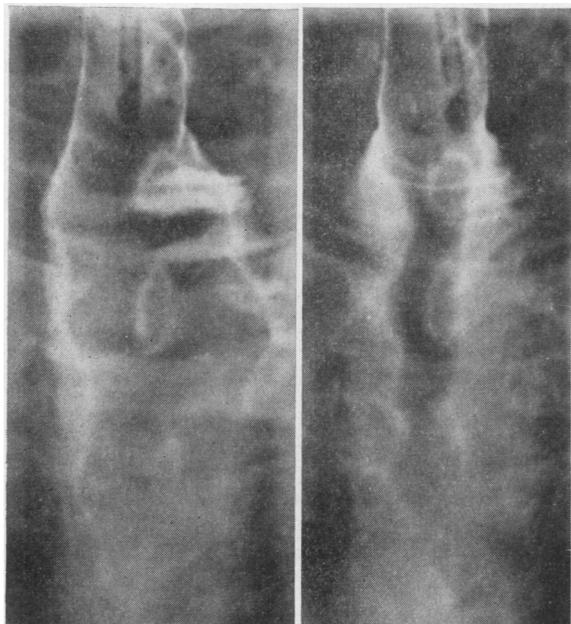


FIG. 5. Case 2. Bronchogram confirms the tracheobronchomegaly and shows the normal segmental and subsegmental bronchi.



FIGS 6 and 7. Case 2. Tracheograms illustrating the massive dilatation of the trachea during inspiration and its collapse during expiration.

The extent of the movement of the trachea is demonstrated in case 2, and bronchiectasis co-exists in case 1. The measurement of the airways diameter is compared with previously reported cases (Table I) and with a control group (Table II).

The milder degree of enlargement of the trachea often seen associated with pulmonary fibrosis and chronic respiratory infection (Shanks and Kerley, 1962), in which only the membranous part of the tracheal wall is affected, should be distinguished from this entity of tracheobronchomegaly in which the marked dilatation of the wall involves both the cartilaginous rings and the membranous part of the wall of the airways, producing the diagnostic bronchographic appearance of a strikingly wide and deeply corrugated tube which gives a false impression of multiple diverticula (Johnston and Green, 1965).

Pathologically, tracheobronchomegaly appears to arise as a congenital defect or atrophy in the connective tissues of the tracheobronchial tree. The few biopsy and necropsy studies of the tracheal wall in this condition show thinning of the muscularis mucosae with paucity of elastic fibres (Rouan, 1959). Brenner and Krauter (1938) observed no evidence of inflammation. The association of tracheobronchomegaly with Ehlers-Danlos syndrome (Aaby and Blake, 1966) may be

further evidence of a connective tissue disorder.

The wide, weak, inefficient tracheobronchial wall predisposes to recurrent respiratory infections, and it is the investigation of these which has led to the diagnosis of most of the reported cases of tracheobronchomegaly. The difficult and ineffective expectoration have been demonstrated clinically and have been confirmed by the bronchoscopic findings (Levowitz, Khalil, Hughes, Conant, and Weiss, 1964).

During bronchography, the contrast medium was shown to be cleared quite normally from the distal bronchi during coughing but to stagnate in the trachea and main stem bronchi (Johnston and Green, 1965).

The undisputed rarity of tracheobronchomegaly makes it unlikely to be an acquired condition secondary to respiratory disease associated with long-standing and productive cough.

Most of the reported cases have been young adults. Doyle, Rellan, and Brea (1954) reported a case in an 8-year-old boy. Tracheobronchomegaly may exist without any associated broncho-

TABLE I
DIAMETERS OF TRACHEA AND BRONCHI IN REPORTED CASES OF TRACHEOBRONCHOMEGALY

Investigator	Age/Sex	Largest Diameter (mm.)		
		Trachea	Right Main Bronchus	Left Main Bronchus
Mounier-Kuhn (1932)	54 F	29	20	16
Katz <i>et al.</i> (1962)	35 M	40	26	30
	39 M	45	32	25
	39 F	35	28	25
	33 M	36	25	20
Johnston and Green (1965)	49 M	43	34	31
	30 M	40	28	30
	35 F	32	29	27
Surprenant and O'Loughlin (1966)	28 M	32	23	15
	42 M	31	25	24
Present report (1967)	24 M	46	28	32
	44 M	40	32	24

TABLE II
DIAMETERS OF TRACHEA AND BRONCHI IN OUR CASES COMPARED WITH THOSE OF 50 CONSECUTIVE BRONCHOGRAMS OF ADULTS (KATZ *et al.*, 1962)¹

	Mean (mm.)	Mean plus Three Standard Deviations	Case 1	Case 2
Trachea	20.2	30.5	46	40
R. main bronchus	16.0	24.0	28	32
L. main bronchus	14.5	23.0	32	24

¹ Measurements made at the largest diameter of the trachea and bronchi

pulmonary disease (Surprenant and O'Loughlin, 1966). Howland, Curry, and Dickinson (1964) reported a case with eight years' follow-up in which no more dilatation of the trachea and main stem bronchi occurred over the years despite progressively severe tracheobronchial infection.

An asymptomatic case was diagnosed in a family survey, and Johnston *et al.* (1965) gave evidence suggesting that tracheobronchomegaly is genetically determined by autosomal recessive inheritance.

CONCLUSION

The rare syndrome of tracheobronchomegaly should be suspected in a patient having an extraordinarily loud, harsh, rasping, and prolonged cough associated with ineffective expectoration.

The unusual width of the trachea may not be shown clearly or may be overlooked in the conventional postero-anterior chest films. The bronchographic findings are diagnostic and may demonstrate associated bronchiectasis.

In future, more complete studies, including histopathology of any recognized case, would be valuable.

We are greatly indebted to Dr. H. M. Foreman, the Physician Superintendent, Sully Hospital, for permission to report the second case and for his valuable help, criticism, and advice.

REFERENCES

- Aaby, G. V., and Blake, H. A. (1966). Tracheobronchiomegaly. *Ann. thorac. Surg.*, **2**, 64.
- Brenner, F., and Krauter, S. (1938). Ueber einen Fall von besonderer Grösse der Luftröhre. *Wien. klin. Wschr.*, **51**, 896.
- Doyle, J. A., Rellan, L. M., and Brea, M. M. (1954). Megatraquea y bronquiectasias congenitas (Síndrome de Mounier-Kuhn). *Rev. Asoc. méd. argent.*, **68**, 579.
- Howland, W. J., Curry, J. L., and Dickinson, D. W. (1964). Tracheobronchiectasis; Report of a case with eight year interval studies. *Dis. Chest*, **45**, 558.
- Johnston, R. F., and Green, R. A. (1965). Tracheobronchiomegaly. Report of five cases and demonstration of familial occurrence. *Amer. Rev. resp. Dis.*, **91**, 35.
- Katz, I., LeVine, M., and Herman, P. (1962). Tracheobronchiomegaly. *Amer. J. Roentgenol.*, **88**, 1084.
- Levowitz, B. S., Khalil, G., Hughes, R. E., Conant, J. S., and Weiss, M. (1964). Tracheobronchiomegaly and chronic pulmonary disease. Report of two cases. *Dis. Chest*, **46**, 498.
- Mounier-Kuhn, P. (1932). Dilatation de la trachée, constatations radiographiques et bronchoscopiques. *Lyon m'éd.*, **150**, 106.
- Rouan, M. (1959). Un cas de trachéomégalie. *J. franç. Méd. Chir. Thor.*, **13**, 417.
- Shanks, S. C., and Kerley, P. (1962). *A Text-Book of X-ray Diagnosis*, 3rd ed., Vol. 2. Lewis, London.
- Surprenant, E. L., and O'Loughlin, B. J. (1966). Tracheal diverticula and tracheobronchomegaly. *Dis. Chest*, **49**, 345.