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Lung function in patients with tracheobronchopathia osteochondroplastica

H. Tukiainen, M. Torkko, E.O. Terho

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ABSTRACT: We have analysed the ventilatory function of seven patients with tracheobronchopathia osteochondroplastica. One patient showed reversible airflow obstruction meeting the criteria of bronchial asthma. Another two patients yielded reduced percentage forced expiratory volume (FEV%) and features of small airways obstruction. The patients' previous spirometric measurements also made the rough estimation of longitudinal changes in lung function possible during a follow-up period of 1–8 yrs (mean 4.2 yrs). Although most patients had suffered severe chest infections, no deterioration in spirometric parameters was found during the follow-up. These findings suggest that tracheobronchopathia osteochondroplastica usually has a benign course.

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Department of Pulmonary Diseases, Kuopio University Central Hospital, Kuopio, Finland.

Correspondence: Dr. H. Tukiainen, Department of Pulmonary Diseases, Kuopio University Central Hospital, SF-71800 Siilinjärvi, Finland.

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Tracheobronchopathia osteochondroplastica is a rare disorder of the tracheobronchial tree. It is characterized by ossified mucosal nodules which protrude into the lumen of the trachea and bronchi. Several theories on the aetiology of this disorder have been proposed, but none has been confirmed [1–3]. So far, over 300 cases have been described in the literature [4]. The syndrome can be detected by bronchoscopy, or may be an incidental finding at autopsy. The patients are often asymptomatic, but various respiratory symptoms may occur. Tracheobronchopathia osteochondroplastica is usually regarded as a benign condition.

The spirometric parameters of ventilatory function can be normal, or a slight intra- or extrathoracic obstruction of the airways may be present [4–10]. However, the data concerning ventilatory function and follow-up of these patients are at present limited. We have analysed the longitudinal changes in ventilatory function of seven patients seen at our clinic during the years 1971–1985.

Patients and methods

The mean age of the patients at the time of ventilatory function measurements was 43.1 yrs, and the mean age at the time when the diagnosis was formed was 39.3 yrs. In one patient the disorder was diagnosed at the age of 20 yrs and in another at the age of 21 yrs. Both of these patients had already experienced respiratory symptoms for many years. The patients' characteristics, medical history and specific respiratory symptoms are summarized in table 1.

Chest X-ray of one patient yielded minor inactive tuberculous sequelae. Otherwise, chest X-rays were normal in all patients except for a costal anomaly found in the chest X-ray of one. Narrowing of the tracheal lumen in routine chest X-ray was suspected in one patient only. In another patient bronchography revealed narrowing of the trachea and irregularities in the contour of the trachea and bronchi.

On bronchoscopy all patients showed the typical mucosal appearance of tracheobronchopathia osteochondroplastica. The changes involved the trachea and/or main or lobar bronchi. In one patient (case 6) the changes extended even into the segmental bronchi. Histopathologic verification of the disorder was obtained in all cases.

Airway resistance, flow-volume curves and static lung volumes were measured, using a body plethysmograph (Gould Autobox 2800). The mean value of three acceptable recordings was adopted in the analyses of airway resistance and static lung volumes. Spirometric indices were analysed according to the recommendations presented by the American Thoracic Society [11]. The measurements were repeated following two puffs of isoprenaline.

Pulmonary diffusing capacity was assessed with a single breath method (Hewlett Packard 47804 S System). The best of three acceptable measurements was recorded (two best measurements not differing more than 10% from each other).

A single breath N_2-washout test (Hewlett Packard 47804 S System) was used for the measurement of closing volume and of the slope of the alveolar plateau (ΔN_2). The mean values from the best two curves were recorded.

In the follow-up of lung function, previous spirometric recordings were used. Original curves were checked, and only curves which fulfilled the criteria presented by the American Thoracic Society [11] were

Table 1. – Clinical data of seven patients with tracheobronchopathia osteochondroplastica

Male/female	4/3
Mean age, yrs (at the time of the study)	43.1 (range 23–59)
Mean age, yrs (at the time of making the diagnosis)	39.3 (range 20–58)
Smoker	0
Ex-smoker	1
Non-smoker	6
Recurrent maxillary sinusitis or ozaena	4 (3 operated)
Chronic rhinitis	6
Recurrent cough or expectoration	6
Haemoptysis	3
Pneumonia	5
Dyspnoea	4
Positive skin prick tests*	2/5
Bronchial hyperreactivity (methacholine inhalation challenge)	0/5
Blood eosinophilia (>5%)	0
Histologic verification	7

*At least two positive reactions (wheal diameter ≥3x3 mm) to common allergens.

accepted. The follow-up period ranged from 1.0–8.5 yrs (mean 4.2 yrs). The apparatus used in the first and final spirometry was not necessarily the same.

Results

The lung function measurements yielded quite normal results (table 2). One patient (case 6) had significant airflow obstruction which involved both large and small airways. On bronchoscopy, this patient had extensive changes of tracheobronchopathia osteochondroplastica extending as far as the segmental bronchi. This patient had, however, also shown reversible airways obstruction on several occasions, and so fulfilled the criteria for bronchial asthma [12]. Two other patients (cases 2 and 5) also displayed a reduced percentage forced expiratory volume (FEV%) and features of small airways obstruction. However, in these patients the airway resistances were within normal limits as well as the indices of inspiratory flow-volume curves. In other patients there was no evidence of significant impairment of ventilatory function. The inspiratory flow-volume curves were also normal, thus excluding any significant extrathoracic airways obstruction. Following isoprenaline inhalation, a bronchodilating effect was observed only in the patient who had previously shown reversible airways obstruction. Pulmonary diffusing capacity was normal in all patients.

The retrospective comparison of spirometric recordings (forced expiratory volume in one second (FEV_1), forced vital capacity (FVC) and FEV%) revealed no deterioration during the follow-up (table 3), except for the patient with reversible airways obstruction. Her spirometric parameters at the final assessment were lower than they had been 8.5 yrs previously.

Table 2. – Individual data of pulmonary function in seven patients with tracheobronchopathia osteochondroplastica

Parameter	Unit	Case 1		Case 2		Case 3		Case 4		Case 5		Case 6		Case 7	
VC	*l**	6.23	(133)	5.07	(109)	5.47	(101)	4.67	(124)	4.97	(112)	2.82	(81)	4.12	(112)
TLC	*l**	8.05	(129)	6.71	(96)	6.97	(95)	6.13	(109)	7.43	(109)	5.37	(99)	5.26	(103)
RV	*l**	1.63	(90)	1.86	(86)	1.42	(80)	1.70	(87)	2.63	(121)	2.47	(143)	1.03	(64)
FVC	*l**	6.30	(134)	5.07	(109)	5.45	(100)	4.67	(124)	4.89	(110)	2.62	(75)	4.08	(111)
FEV_1	*l**	4.67	(128)	3.40	(90)	5.00	(105)	3.77	(130)	3.16	(88)	1.73	(61)	3.31	(114)
FEV	%*	74.0	(77)	67.0	(81)	91.0	(88)	81.0	(76)	64.0	(81)	66.0	(81)	81.0	(78)
FIV_1	*l*	4.46		4.41		5.22		3.47		3.96		1.69		3.73	
FIV	%	76.0		92.0		95.0		78.0		82.0		62.0		90.0	
FEF_{50}	% $l{\cdot}s^{-1}$	4.80	(103)	2.55	(51)	7.02	(104)	4.78	(121)	2.90	(59)	1.24	(30)	4.06	(104)
Raw	$kPa{\cdot}l^{-1}{\cdot}s^{-1}$*	0.136	(126)	0.146	(107)	0.101	(81)	0.108	(109)	0.086	(66)	0.382	(224)	0.117	(113)
sGaw	$kPa^{-1}{\cdot}s^{-1}$*	1.85	(79)	2.17	(109)	3.27	(157)	2.63	(96)	2.48	(124)	0.75	(43)	2.54	(93)
CV/VC	%**	10.0	(13.7)	34.1	(20.5)	9.1	(7.3)	12.8	(15.5)	39.6	(21.7)	13.8	(18.3)	12.4	(7.5)
ΔN_2**		0.71	(84)	1.53	(176)	0.64	(77)	0.89	(79)	2.38	(274)	4.01	(397)	0.85	(77)
DLCO	$mmol{\cdot}min^{-1}{\cdot}kPa^{-1}$*	11.4	(141)	10.7	(127)	15.0	(147)	8.4	(111)	7.4	(92)	6.2	(85)	9.2	(126)

*: Reference values according to VILJANEN [18]; **: Reference values according to BUIST *et al.* [19]. Percentage of predicted values are expressed in parenthesis except for FEV%, and CV/VC% for which predicted values are shown. VC: vital capacity; TLC: total lung capacity; RV: residual volume; FVC: forced vital capacity; FEV_1: forced expiratory volume in one second; FIV_1: forced inspiratory volume in one second; FEF_{50}%: forced mid-expiratory flow; Raw: airways resistance; sGaw: specific airways conductance; CV/VC%: closing volume as percentage of vital capacity; DLCO: diffusing capacity of carbon monoxide.

Table 3. – Retrospective comparison of lung function (FEV_1 and FVC) of seven patients with tracheobronchopathia osteochondroplastica.

	First assessment		Second assessment		Time interval
	FEV_1 *l*	FVC *l*	FEV_1 *l*	FVC *l*	yrs
Case 1	4.19	5.84	4.67	6.30	0.8
Case 2	2.95	4.59	3.40	5.07	2.3
Case 3	4.89	5.36	5.00	5.45	3.6
Case 4	3.75	4.50	3.77	4.67	5.3
Case 5	3.30	5.30	3.16	4.89	0.8
Case 6	2.65	3.85	1.73	2.62	8.4
Case 7	3.25	3.50	3.31	4.08	8.4
Mean	3.57	4.71	3.58	4.73	4.2
±SD	±0.77	±0.85	±1.08	±1.15	

FEV_1: forced expiratory volume in one second; FVC: forced vital capacity.

Discussion

The present results are in accordance with the view that progressive deterioration of ventilatory function is not typical for tracheobronchopathia osteochondroplastica. Previously, a significant worsening in ventilatory function during an 8-month follow-up of one patient has been reported by ALROY *et al.* [10]. In contrast, NIEROP *et al.* [9] found no progressive deterioration in lung function of one patient over an 8 yr follow-up. The mucosal appearance checked by bronchoscopy also remained unchanged. In addition, no obvious change was observed in bronchial pathology of another patient whose bronchoscopy was repeated with a 1 yr interval [13]. ROSE *et al.* [14] have also reported a case in which the endoscopic view remained unchanged over a 6 yr follow-up.

Chest infections seem to be common in these patients [6–8, 15], and recurrent pneumonia was characteristic in our subjects. However, the occurrence of respiratory infections did not seem to lead to progressive deterioration of lung function. In this respect, tracheobronchopathia osteochondroplastica seems to resemble the productive type of chronic bronchitis in its benign course [16].

One patient in our study showed reversible airflow obstruction. At the moment of control, the spirometric values of this patient may have been low by chance, due to spontaneously changing obstruction of simultaneous asthma. The study group was too small for further conclusions about the relationships between tracheobronchopathia osteochondroplastica and asthma or reversible bronchial obstruction. Features of minor expiratory obstruction were also observed in another two patients. This finding is consistent with previous observations that slight expiratory and/or inspiratory obstruction may occur in some patients. It could be expected that the bony nodules of the tracheal and bronchial mucosa might cause airflow limitation. On the other hand, it may be assumed that the bronchial tree with bony formations is rather stiff, thus opposing tendencies for an intrathoracic airway collapse. The normal diffusing capacity in our study excludes any significant abnormality of the gas exchanging surfaces of the lung.

Tracheobronchopathia osteochondroplastica is not usually diagnosed before the age of 50 yrs. In our study there were two young patients. There are also some other reports about the condition in young people [13, 15, 17]. This also favours the concept that the disorder has a benign course and at least in some patients it may have an early, perhaps even congenital, onset.

There may exist geographical differences in the occurrence of this disorder. For example, according to the British literature, it seems to be extremely rare [13]. In Finland, in addition to our material, HÄRMÄ *et al.* [15] have presented thirty patients. These regional differences may be due to differences in diagnostic practice, but it is also possible that the differences in prevalence are real and due to the different genetic background of populations.

Our analysis of follow-up data was based on previous spirometric recordings. Admittedly this kind of analysis may be subject to severe bias due to possible changes in spirometric methodology. In fact, the spirometric apparatus used in this study was not necessarily the same in the first and the final assessment. In addition, the number of patients was rather small. Taking into account these limitations, an approximate estimation of changes in spirometric parameters is, however, possible. No essential change was found, which, together with the few previous findings, speaks in favour of a benign nature for tracheobronchopathia osteochondroplastica.

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RÉSUMÉ: Nous avons étudié la fonction ventilatoire de 7 patients atteints de trachéobronchopathie ostéochondroplastique. Chez l'un d'entre eux, on a relevé une obstruction réversible des débits aériens rencontrant les critères de l'asthme bronchique. Deux autres patients avaient une diminution du VEMS et des signes d'obstruction des petites voies aériennes. Les mesures spirométriques antérieures, réalisées chez ces patients, ont permis d'étudier les modifications longitudinales de la fonction pulmonaire au cours d'un follow-up d'un à huit ans (moyenne 4.2). Quoique la plupart des patients aient eu des infections bronchiques sévères, l'on n'a pas trouvé de détérioration des paramètres spirométriques pendant le suivi. Ces constatations suggèrent que la trachéobronchopathie ostéochondroplastique a habituellement un décours bénin.