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CORRESPONDENCE

An 86-yr-old female with lymphangioleiomyomatosis

To the Editors:

In the paper by JOHNSON [1] on lymphangioleiomyomatosis, he describes the typical patient being female and of child-bearing age. Although patients outside this age range have been described, the oldest recorded in the recently reported National Heart, Lung and Blood Institute registry was aged 76 yrs, the mean age of onset being 38.9 yrs [2].

We wish to report a case of lymphangioleiomyomatosis in an 86-yr-old female who presented with acute breathlessness on a background of increasing dyspnoea over a 3-yr period. She had been told that she had chronic obstructive pulmonary disease 2 yrs prior to this presentation, despite having never smoked. At the time of admission she was taking regular nebulised salbutamol but no other respiratory medications. She had previously undergone resective surgery for breast carcinoma and was on continuing treatment with anastrazole. Several years previously, she had also undergone a hysterectomy and bilateral oopherectomy, but was unclear as to why. There was no other significant medical history. On examination, she was comfortable at rest, but tachypnoeic and hypoxic on minimal exertion (oxygen saturation of 88% on room air). Apart from hyperinflation, the rest of the chest examination was unremarkable. Blood investigations including α_1-antitrypsin assays were within normal limits. A computed tomography pulmonary angiogram was arranged as there was concern that she had suffered a pulmonary embolus. No emboli were noted; however, diffuse bilateral thin-walled cysts were identified. Two chest radiologists each reviewed the scans independently and concluded that these appearances were most consistent with the diagnosis of lymphangioleiomyomatosis (fig. 1). We speculate that the patient's previous oophorectomy and use of the aromatase inhibitor anastrazole had ameliorated the condition, which led to its late presentation. Although a rare condition, we suggest lymphangioleiomyomatosis be considered in the differential diagnosis of dyspnoea in elderly females who demonstrate atypical features of airflow limitation in their presentation.

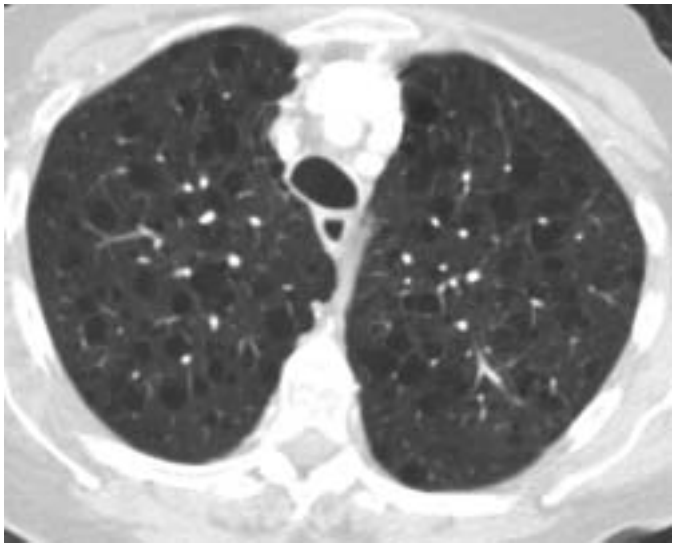

FIGURE 1. Computed tomography scan of the chest revealing multiple thin-walled cysts, typical of lymphangioleiomyomatosis.

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Cardiorespiratory screening for sleep-disordered breathing

To the Editor:

Sleep-disordered breathing (SDB) is associated with an increasing mortality [1, 2]. The prevalence of SDB, in particular central sleep apnoea and Cheyne–Stokes respiration, is remarkably high in heart failure patients [3]. Therefore, screening for SDB in heart failure patients is an emerging clinical problem.

Waiting times for in-hospital polysomnography (PSG), which still represents the gold standard for SDB diagnosis, are

increasing. Consequently, cardiorespiratory polygraphy (PG) devices for the diagnosis of SDB have been introduced. Two studies of PG devices have previously been published in the *European Respiratory Journal*. DINGLI *et al.* [4] compared data of PSG with those obtained with a portable PG device (Embletta; Medcare, Reykjavik, Iceland) to detect obstructive sleep apnoea. Using simultaneous measurements they found a close agreement in total apnoea/hypopnoea index (AHI) scores ($29.2\pm3.7 \cdot h^{-1}$ in PSG *versus* $27.2\pm3.4 \cdot h^{-1}$ in PG). QUINTANA-GALLEGO *et al.* [5] assessed the benefit of ambulatory PG (Apnoescreen II; Erich Jaeger, Wuerzburg, Germany) to establish the diagnosis of SDB in heart failure patients. They reported a close correlation in the results of PSG *versus* PG measurements with a high sensitivity and specificity. In both studies, recordings were reviewed by sleep specialists.

Analysing software for PG recordings may further contribute to an increase in the number of screened patients. The use may be suitable especially in high volume cardiology centres with large numbers of heart failure patients. We used the Embletta device as introduced by DINGLI *et al.* [4] to screen SDB in 104 consecutive patients. Somnologica for Embletta (Version 3.3; Medcare) was used for a first ever analysis (automatic analysis). All recordings were then reviewed by two sleep specialists (manual analysis), blinded to the automatic analysis results. Default settings and standard definitions were used for the detection of apnoea (complete cessation of airflow for $\geqslant$10 s) and hypopnoea ($\geqslant$50% reduction in respiratory airflow accompanied by a decrease of $\geqslant$4% in arterial oxygen saturation lasting for 10 s).

For the most widely used parameter in SDB, the AHI, the correlation between automatic and manual analysis is shown (fig. 1); as well as sensitivity, specificity, and positive and negative predictive values (table 1) for several AHI cut-off values. In addition, a Bland–Altman plot is presented (fig. 2), as recommended by FLEMONS and LITTNER [6].

In addition to the results of DINGLI *et al.* [4] and QUINTANA-GALLEGO *et al.* [5], the current data show that automatic analysis of PG recordings is effective for ruling out sleep apnoea, with a 96.4% specificity, when using the clinically relevant AHI cut-off of $\geqslant15 \cdot h^{-1}$. However, the sensitivity of 72.9% with automatic (not reviewed) analysis lead to a relevant underestimation of SDB in ~25% of patients. Somnologica for Embletta software tended to underscore AHI, when compared with manual review by two independent sleep specialists.

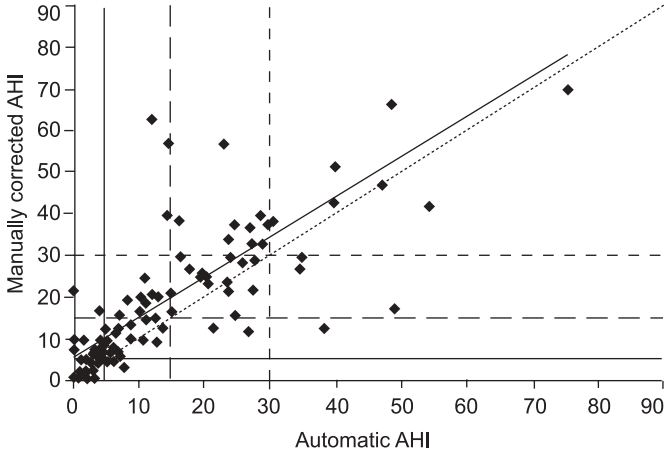

FIGURE 1. Correlation of automatic *versus* manually obtained apnoea/hypopnoea index (AHI) in 104 consecutive patients screened for the presence of sleep-disordered breathing. ——: AHI $\geqslant5 \cdot h^{-1}$; – – –: AHI $\geqslant15 \cdot h^{-1}$; - - -: AHI $\geqslant30 \cdot h^{-1}$; y=0.9685x+4.8129; r^2=0.6664.

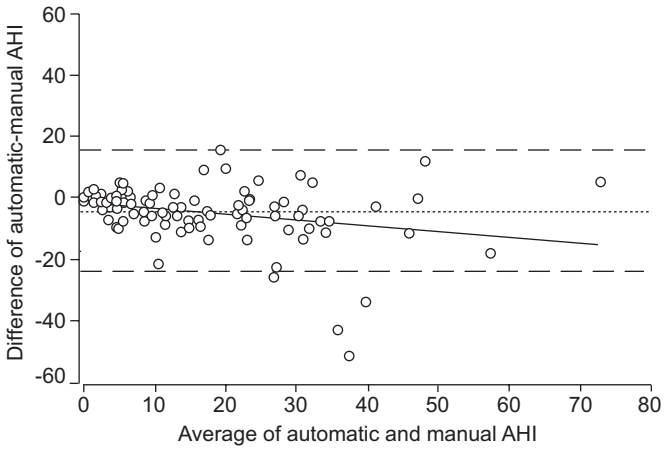

FIGURE 2. Bland–Altman plot of mean differences and limits of automatic *versus* manually obtained apnoea/hypopnoea index (AHI) values in 104 consecutive patients screened for sleep-disordered breathing. The coefficient of variation (2 SD) is 18.54 events·h^{-1}, with a trend toward increased error at larger AHI levels. ·····: mean difference; – – –: $\pm$2SD; y=-0.1876×-1.4248; r^2=0.0812.

In agreement with QUINTANA-GALLEGO *et al.* [5] we believe that cardiorespiratory polygraphy is an adequate tool for cardiology screening, especially heart failure patients, for the presence, type and severity of sleep-disordered breathing, but recordings have to be carefully reviewed by specially trained personnel.

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TABLE 1 Sensitivity, specificity, and negative and positive predictive value of automatically obtained apnoea/hypopnoea index (AHI) values at several cut-offs[#]

	Sensitivity	Specificity	Predictive value	
			Positive	Negative
AHI $\geqslant5 \cdot h^{-1}$	0.780	0.864	0.955	0.514
AHI $\geqslant15 \cdot h^{-1}$	0.729	0.964	0.946	0.806
AHI $\geqslant30 \cdot h^{-1}$	0.368	0.976	0.778	0.874

[#]: A high specificity means that the analysis performed is good for ruling out sleep apnoea at the expense of decreased sensitivity.

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Prejudgement towards the quality of spirometry in primary care does not help our case

To the Editors:

We very much appreciate the contribution by ZIELINSKI *et al.* [1] on the topic of early detection of chronic obstructive pulmonary disease (COPD) by high-risk population screening in the April issue of the *European Respiratory Journal*. The authors mention that ''insufficient quality of spirometries performed in the primary care setting was also reported recently by SCHERMER et al.'' We were quite surprised to notice that in referring to our paper on the validity of spirometry in general practice [2], ZIELINSKI *et al.* [1] fail to appreciate the observation that the 61 primary-care practices involved in our study actually achieved comparable, or even slightly higher, forced expiratory volume in one second and forced vital capacity values, and produced an identical proportion of reproducible tests as the four hospital-based pulmonary function laboratories involved. In contrast to our findings, the study reported recently by ENRIGHT *et al.* [3] clearly demonstrates that the performance rate of technicians in a pulmonary function laboratory can indeed be very high. However, significant variation in spirometry test results between laboratories has previously been reported by DOWSON *et al.* [4]. This should be kept in mind when judging the quality of spirometry in primary-care settings.

In addition, we have previously shown that trained primary-care physicians are able to recognise obstructive flow patterns correctly in >90% of cases [5]. Sufficient quality of basic spirometry tests and recognition of subjects with undiagnosed COPD can thus be performed in the primary-care setting, provided that personnel is adequately trained and regular feedback on performance is available. When touching upon the important issue of raising COPD awareness, one ought to consider mobilising the vast potential of primary healthcare professionals, who deliver essential care to the majority of patients with chronic respiratory disease worldwide.

The suboptimal accuracy of primary-care spirometry may come at the cost of an inevitable but small number of false-positive cases. Still, when one pursues to reach the large pool of as yet undiagnosed subjects with chronic obstructive pulmonary disease in the population, the involvement of primary care professionals is indispensable.

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