

Form the authors:

We thank G.L. Casoni and V. Poletti for their comments regarding our study showing the diagnostic utility of a thin bronchoscope with a 1.7-mm working channel for peripheral pulmonary lesions [1]. Their main quarrel with a channel of this size is its possible inability to control bleeding, and they argue that larger forceps are better than smaller ones in terms of diagnostic yield.

Bleeding is one of the well-known and potentially serious complications associated with transbronchial biopsy (TBB). We think that the risk of severe bleeding associated with thin bronchoscopy is lower than with conventional bronchoscopy. Biopsy with small forceps is likely to reduce the risk of a massive haemorrhage. Additionally, the technique, in which the tip of the thin bronchoscope is wedged firmly into the peripheral bronchi during TBB, may also contribute to the reduction of bleeding complications. Of course, the safety of the procedure was one of the key points in the evaluations in our study, and no significant bleeding which could not be controlled by the thin bronchoscope was observed. We think this procedure is indeed safe on the basis of the results.

The second issue raised is controversial, whether or not the size of the biopsy forceps influences the diagnostic yield [2, 3]. The British Thoracic Society Bronchoscopy Guidelines Committee [4] noted that "the type of forceps used does not seem to influence the diagnostic yield." Although the results of the published studies concerning the relationship between the diagnostic yield and the size of forceps might not be applicable to the small forceps for a 1.7-mm working channel, several studies [5, 6] using these forceps, including ours, have demonstrated field-proven results in terms of diagnostic yield.

We agree that endobronchial ultrasound is a useful adjunct to bronchoscopy. Other new modalities such as electromagnetic navigation, virtual bronchoscopic navigation or computed tomography fluoroscopy should also be useful as well as thin bronchoscopy. These new modalities seem to offer improved diagnostic yield compared with conventional bronchoscopy,

although none of them may be optimal. In any case, we bronchoscopists must not be content with the "best approach," which is only the best approach so far. There is indeed a better way to diagnose peripheral pulmonary lesions; we simply must find it.

M. Oki and H. Saka

Dept of Respiratory Medicine, Nagoya Medical Center, Nagoya, Japan.

STATEMENT OF INTEREST

None declared.

REFERENCES

- 1 Oki M, Saka H, Kitagawa C, *et al.* Novel thin bronchoscope with a 1.7-mm working channel for peripheral pulmonary lesions. *Eur Respir J* 2008; 32: 465–471.
- 2 Wang KP, Wise RA, Terry PB, *et al.* Comparison of standard and large forceps for transbronchial lung biopsy in the diagnosis of lung infiltrates. *Endoscopy* 1980; 12: 151–154.
- 3 Smith LS, Seaquist M, Schillaci RF. Comparison of forceps used for transbronchial lung biopsy: bigger may not be better. *Chest* 1985; 87: 574–576.
- 4 Honeybourne D, Babb J, Bowie P, *et al.* British Thoracic Society guidelines on diagnostic flexible bronchoscopy. *Thorax* 2001; 56: Suppl. 1, i1–i21.
- 5 Yoshikawa M, Sakoh N, Yamazaki K, *et al.* Diagnostic value of endobronchial ultrasonography with a guide sheath for peripheral pulmonary lesions without X-ray fluoroscopy. *Chest* 2007; 131: 1788–1793.
- 6 Yamada N, Yamazaki K, Kurimoto N, *et al.* Factors related to diagnostic yield of transbronchial biopsy using endobronchial ultrasonography with a guide sheath in small peripheral pulmonary lesions. *Chest* 2007; 132: 603–608.

DOI: 10.1183/09031936.00127808

Familial spontaneous pneumothorax: importance of screening for renal tumours

To the Editors:

We read with interest the recent article by FRÖHLICH *et al.* [1], wherein the authors reported two new disease-associated DNA sequence alterations in *FLCN*, the tumour suppressor gene located at chromosome 17p11.2 which, when mutated, leads to the genodermatosis Birt–Hogg–Dubé syndrome (BHDS). However, we feel that there is an important point that deserves greater prominence.

BHDS was named after three Canadian dermatologists who, in 1977, described 15 adults in a kindred of 70 who had multiple small, dome-shaped papular skin lesions, presenting at >25 yrs

of age, over the scalp, forehead, face and neck, with scattered lesions on the chest and back [2]. Histologically, these lesions were confirmed to be fibrofolliculomas, benign hamartomas of the hair follicle. Subsequently, this syndrome was found to be a marker of internal disease, as cases of recurrent pneumothorax, lung cysts [3] and renal tumours [3, 4] were reported.

BHDS has now been recognised as one of the inherited renal cancer syndromes, which include von Hippel Lindau, hereditary papillary renal carcinoma and hereditary leiomyomatosis renal cell carcinoma [5]. Renal tumours have been reported in as many as 34% of individuals with germline *FLCN* mutations [6].

They are frequently multiple and bilateral, and present at a mean age of 50.7 yrs [7]. The most common histological subtypes are hybrid oncocyctic (50%) and chromophobe (34%) renal cell carcinoma, while clear cell, oncocytoma and papillary renal cell cancer are less frequently found [7]. Radiographic screening is recommended, with a typical strategy involving abdominal computed tomography and/or renal ultrasound at the time of diagnosis, followed by interval screening every 3–5 yrs [8]. Parenchyma-sparing surgery is recommended given the risk of development of further tumours [7]. In line with these recommendations, all participants in the study by FRÖHLICH *et al.* [1] were screened for kidney manifestations by abdominal ultrasound and none were identified.

The importance of recognising the possibility of a diagnosis of Birt–Hogg–Dubé syndrome in patients with familial recurrent pneumothorax, with or without characteristic skin findings, lies in undertaking surveillance for renal tumours, a potentially lethal feature of this syndrome.

G. Warwick*, L. Izatt[#] and E. Sawicka[†]

*Dept of Thoracic Medicine, St Vincent's Hospital, Sydney, Australia. [#]Dept of Clinical Genetics, Guys Hospital, London, and [†]Dept of Respiratory Medicine, Princess Royal University Hospital, Orpington, Kent, UK.

STATEMENT OF INTEREST

None declared.

REFERENCES

- 1 Fröhlich BA, Zeitz C, Mátyás G, *et al.* Novel mutations in the folliculin gene associated with spontaneous pneumothorax. *Eur Respir J* 2008; 32: 1316–1320.
- 2 Birt AR, Hogg GR, Dubé WJ. Hereditary multiple fibrofolliculomas with trichodiscomas and acrochordons. *Arch Dermatol* 1977; 113: 1674–1677.
- 3 Toro JR, Glenn G, Duray P, *et al.* Birt-Hogg-Dubé syndrome: a novel marker of kidney neoplasia. *Arch Dermatol* 1999; 135: 1195–1202.
- 4 Roth JS, Rabinowitz AD, Benson M, Grossman ME. Bilateral renal cell carcinoma in the Birt-Hogg-Dubé syndrome. *J Am Acad Dermatol* 1993; 29: 1055–1056.
- 5 Linehan WM, Pinto PA, Srinivasan R, *et al.* Identification of the genes for kidney cancer: opportunity for disease-specific targeted therapeutics. *Clin Cancer Res* 2007; 13: 671s–679s.
- 6 Toro JR, Wei MH, Glenn GM, *et al.* BHD mutations, clinical and molecular genetic investigations of Birt-Hogg-Dubé syndrome: a new series of 50 families and a review of published reports. *J Med Genet* 2008; 45: 321–331.
- 7 Pavlovich CP, Walther MM, Eyler RA, *et al.* Renal tumors in the Birt-Hogg-Dubé syndrome. *Am J Surg Pathol* 2002; 26: 1542–1552.
- 8 Welsch MJ, Kronic A, Medenica MM. Birt-Hogg-Dubé syndrome. *Int J Dermatol* 2005; 44: 668–673.

DOI: 10.1183/09031936.00126608

Impulse oscillometry in comparison to spirometry in pregnant asthmatic females

To the Editors:

Asthma complicates 3–8% of pregnancies [1, 2] and early diagnosis and optimal management of the condition seems necessary. However, spirometry as a clinical measure of airway disease mainly depends on subjects efforts during forced expiratory manoeuvres. Considering the high prevalence of respiratory symptoms and breathlessness in pregnancy and the limited functional capacity and mobility of the diaphragm because of the growing foetus [3], an easier, more rapid screening test that does not require patient cooperation would be ideal. Our study aimed to evaluate the correlation between spirometry and impulse oscillation technique (IOS) parameters to reveal the utility of IOS parameters in diagnosis of airflow obstruction in pregnant females.

In total, 125 pregnant females were categorised in three groups of asthmatics: 1) 40 physician-diagnosed asthmatics, on the basis of National Asthma Education and Prevention Program guidelines [4]; 2) 35 probable asthmatics, with symptoms and signs of asthma but normal spirometry; and 3) 50 healthy controls without any signs and symptoms of asthma and with normal spirometry. After obtaining informed consent, baseline

IOS and spirometry measurement, salbotamul (two puffs, 200 µg) was administered by a metered-dose inhaler *via* a spacer device. All tests were performed again 15 min later.

As predicted, at baseline, forced expiratory volume in one second (FEV₁) was significantly lower in the asthmatic group compared with the probable asthmatics and healthy subjects. Forced vital capacity (FVC) was significantly lower and impedance at 5 Hz (Z₅), resistance at 5 Hz (R₅) and resonant frequency (F_{res}) were significantly higher in asthmatics compared with healthy subjects. Resistance at 20 Hz (R₂₀) did not differ between the three groups.

In the asthmatic group, FEV₁ and FVC significantly increased after bronchodilator use and Z₅, R₅ and R₂₀ significantly decreased after bronchodilator use. In asthmatics and probable asthmatics, there was a 15–20% decrease in Z₅, R₅ and R₂₀ after bronchodilator administration (table 1). However, in some of the healthy subjects a small bronchoconstriction after bronchodilator administration was observed but it was not significant (p>0.05).

To our knowledge, this is the first study to report the comparison of IOS values to spirometric values in pregnant