Exertional haemoptysis: LAM and TSC

Tuberous sclerosis (TSC) is characterised by the occurrence of hamartomas in different organs. It is autosomal dominant with complete penetrance and variable expression. TSC is associated with epilepsy, learning difficulties, behaviour problems, and renal and dermatological pathology. Lymphangioleiomyomatosis (LAM) is principally a pulmonary condition characterised by smooth muscle (leiomyo) proliferation around lymphatics (lymph), blood vessels (angio), and alveolar airways. Cystic destruction of lung parenchyma results in the development of pneumothorax. 50% of patients with LAM have renal angiomylipomas which are also the chyma results in the development of pneumothorax. There was a single subungual fibroma. Cardiovascular and respiratory examinations were normal. Pulmonary function tests showed normal lung volumes: FEV, 2.72 l, FVC 3.43 l, TLC 5.21 l, and RV 1.96 l with a corrected transfer factor of 73% predicted. Bronchoscopic examination revealed no source of bleeding. A high resolution CT scan of the thorax showed multiple cystic spaces with well defined walls and normal intervening lung (fig 1). A contrast CT scan of the head showed a single densely calcified subependymal nodule related to the right lateral ventricle. An abdominal CT scan identified multiple renal lesions bilaterally and a single hepatic lesion. Renal biopsy confirmed the presence of angioleiomyomas. The above findings fulfilled the criteria for a diagnosis of LAM and TSC.1 In view of the diverse clinical course of LAM and the questionable value of hormone therapy, the patient was not commenced on treatment but referred for genetic screening.2 This case underscores the need to consider such a diagnosis in female patients presenting with solitary exertional haemoptysis.

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References

Dysfunctional breathing in COPD

I was interested to read Dr Morgan’s review of dysfunctional breathing in asthma in the 2002 Year in Review,3 but the problem may be even greater in COPD. Dr Morgan suggests that the problem may have serious consequences in terms of mortality, but we have published indirect evidence of an association with mortality. In the 10 year follow up of the Darlington and Northallerton Asthma Study the odds ratio for the risk of dying in those who had no best function recorded was 2.5, equivalent to a risk of best function of 60% predicted.4 Although failure to obtain best function was sometimes associated with steroid phobia, by far the most frequent cause was an inability to complete spirometric tests which is a sensitive indicator of dysfunctional breathing.

In non-clinical practice one sees large numbers of patients managed in primary care who have breathlessness attributed to COPD which may or may not exist objectively. By the time they are seen the subjects usually are genuinely breathless because of deconditioning. There is an urgent need to correct this under recognition of the problem. Perhaps a change in the approach to history taking might be helpful. Breathlessness is usually regarded not only as a symptom of COPD—which it may be—but also as a measure of disability due to physiological limitation—which it certainly is not in moderate airway obstruction. The prime measure of disability in chronic cardiorespiratory dysfunction is exercise limitation. If this is physiologically

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Figure 1 From pulmonary high resolution CT scan showing multiple cystic regions with normal intervening lung parenchyma.
mediated through failure of oxygen delivery, then the natural limiting symptom is muscle failure and not breathlessness. This is well recognised in athletes, where breathlessness is accepted as incidental. In as much as breathlessness is due to moderate airway obstruction, it is a mechanical in origin and should be regarded as a contributory factor to exercise limitation rather than its prime cause. Moreover, breathlessness is the initi- ator of the vicious circle of decreased physical activity, deconditioning, and breathlessness which leads to the prime cause of exercise limitation deconditioning. A shift in history taking first to establish the extent of exercise limitation and then to ask about the associ- ated symptoms would lead to a much better approach to the management of chronic respiratory disease, particularly in patients with other chronic diseases that themselves lead to exercise limitation. Perhaps respiratory physicians should train themselves to intro- duce breathlessness last rather than first when talking to a patient.

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Reference

Occupational asthma evaluation
We read with interest the paper by Baldwin et al on the level of agreement between expert clinicians and OASYS software when making a diagnosis of occupational asthma. Our clinical unit uses OASYS plotting regularly, and finds it of great use as one element of the diagnostic toolkit available for the confirmation of a diagnosis of occupational asthma. We were interested to note that there was a low level of agreement between experts and OASYS when peak expiratory flow (PEF) records were interpreted, but agreement within experts was better. We would be inter- ested to know whether the information provided to the experts on the nature of the work was used in determining their final outcome—that is, if an individual was working with a known sensitiser or was in a perceived high risk job, did this influence the outcome more than the graphical and math- ematical data?

In the clinical setting a decision is made to perform regular PEF monitoring in those patients who are thought to have a reasonable situation of seeing patients in secondary care following a prolonged period of sickness absence, making diagnosis even more challenging.

At present the consistency of diagnosis of occupational asthma throughout the UK is likely to be highly variable. We are currently involved in a multicentre UK based study assessing the application of the toolkit to diagnose occupational asthma, and it is evident that practice and results are disparate between various expert centres.

We are sure that the future of occupational asthma evaluation will and should rely on programs like OASYS, but that the diagnosis must be seen also in broader terms, taking into account clinical, immunological, and exposure data.

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Author’s reply
Experts were deemed to “under report” possible cases of occupational asthma. While this may indeed be the case, an alternative explanation is that the experts were more realistic, taking into account the clinical like- lihood as well as the PEF pattern. OASYS sys- tems clearly invoke complex comparisons between known cases of occupational asthma and the record being assessed. The authors suggest that PEF interpretation is best left to experts. While we agree that expert centres which consistently diagnose occupational asthma are needed, as many as one in 10 adult asthmatic patients is likely to have a substantial effect from work. It is therefore important for all such patients in the UK to have access to competent individu- als trained to assess these patients. This is where OASYS (or similar) systems are likely to be very important as an initial screen, and could be carried out by primary care or occupa- tional health nurses or other competent non-clinical people in the workplace. This would enable patients currently working to undergo PEF assessment, as opposed to the common situation of seeing patients in secondary care following a prolonged period of sickness absence, making diagnosis even more challenging.

We are sure that the future of occupational asthma evaluation will and should rely on programs like OASYS, but that the diagnosis must be seen also in broader terms, taking into account clinical, immunological, and exposure data.

We hope we have provided a tool for use by the non-expert in the initial assessment of occupational asthma. We agree that these records need to be made as soon as the diag- nosis is suspected and before workers are removed from their jobs. Supervising such work does, however, require a lot of expertise with particular emphasis on record- ing working times, keeping treatment con- stant, and recording the timings of readings. Help is provided for this on the website occupationalasthma.com, as well as suitable record forms with instructions which can be downloaded.

Ideally, OASYS should be used interactively. The patient returns to clinic with his or her record stored in an electronic meter. The clini- cian and patient review the record together. This allows the clinician to ask those ques- tions suggested by the record such as “Did you have a respiratory infection last week?” (if there was an unexpected fall in PEF crossing work/rest interfaces), or “Remind me of your work pattern on the 25th of last month?” (when a single work day shows no deteriora- tion where others do). The integration of clini- cal information and record is thus even closer, enhancing the diagnostic toolkit referred to by Dr Fishwick and colleagues.

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Lung function in preschool children
We read with great interest the recent paper by Nystad et al on the feasibility of spirometric tests in preschool children using candle blowing incentives, in support of recent publications.1,2 As there is a dearth of spirome- tric reference data for this age group, we value the additional regression equations derived. However, we have several questions concerning this study.

The regression formulae presented were based on 603 children, of which 476 (78.9%) were reported as having “asthmatic symp- toms” or “parental smoking habits”. It would be interesting to stratify the results, analysing healthy and non-healthy populations sepa- rately.

The actual age distribution of the preschool population in table 1 ranged from 4.3 to 4.8 years (that is, age 4 years). This narrow age distribution may explain the high r values of the linear regressions shown in table 4. Evalu- ating younger and older children may de- crease the r values of logarithmic regression. Linear regressions should be used cautiously since parameters may appear to be too low in older children and “negative” in those who asthmatur (fig 1). The “candle blowing” incentives were as- sumed to facilitate technically correct spirome- tric tests in the young children. We found that such incentives induced premature ter- mination of forced vital capacity (FVC) which led to lower values than with other methods.3,4 If this is not the case, how do the authors explain the lower FVC values com- pared with those of Eigen et al, while the forced expiratory volume in 1 second (FEV1) values were similar (fig 3)?  Acceptance criteria for correct FVC curves are vague in the absence of expiration time and “end of test” criteria.5 Inclusion of curves with a difference of 10% between the

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two best curves should be avoided on the basis of standard recommendations and previously published data (≤5% difference only). In view of the increasing interest in lung function in preschool children, resolving these questions would help to standardise spirometric parameters in this age group.

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References

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