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, behavioural 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 pneumothoraces. 50% of patients with LAM have angiomylipomas which are also the most common renal lesion in TSC.1

On examination the patient had a rash on the chin which reportedly bled following viral infections. There was a single subungual fibroma. Cardiovascular and respiratory examinations were normal. Pulmonary function tests showed normal lung volumes: FEV1, 2.72 l, FVC 3.43 l, TLC 5.21 l, and RV 1.96 l with a corrected transfer factor of 73% predicted. Bronchoscopy 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 angiomylipomas.

The above findings fulfil 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.3 This case underscores the need to consider such a diagnosis in female patients presenting with solitary exertional haemoptysis.

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Diaphragm plication following phrenic nerve injury

We read with great interest the paper by Simansky et al describing the good results of plication of the diaphragm following phrenic nerve injury. The authors conclude that pulmonary function tests (PFTs) in combination with quantitative perfusion scans are helpful in selecting patients for this procedure. In table 4 they present the PFTs they were using and, in addition, they suggest that more sophisticated tests such as ultrasoundography or fluoroscopy can also be useful in assessing diaphragmatic paralysis. Although we agree that all these tests are very helpful, assessment of vital capacity (VC) in both sitting and supine positions was omitted. This is a very simple test that gives important information about the function of the diaphragm, with a decrease in VC of >30% from the sitting to the supine position suggesting diaphragmatic paralysis.

The practical value of this test is clearly shown in the following patient in whom we initiated non-invasive positive pressure ventilation (NIPPV) because of a right sided diaphragmatic paralysis due to a coronary bypass. At the start of NIPPV there was a gap between the VC in the sitting and supine positions of 0.8 l (30%), VC sitting 2.7 l, VC supine 1.9 l. We started NIPPV and the patient became less dyspnoeic and less tired. After 18 months the clinical situation was still improving, with an increase in VC both in the sitting and supine positions to 3.5 l and 2.8 l, respectively. After 36 months the gap between VC in the two positions had almost disappeared (3.6 l and 3.5 l, respectively). In addition, the radiograph of the thorax showed a downward shift and normalisation of the position of the right diaphragm. We therefore stopped NIPPV and after several weeks the patient slept well without ventilatory support. This case illustrates that the assessment of VC in both the sitting and supine positions can be very helpful in the diagnosis and follow up of patients with diaphragmatic paralysis.

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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, but the problem may be even greater in COPD.

Dr Morgan suggests that the problem may have serious consequences in terms of morbidity, 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.2 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 cardiopulmonary dysfunction is exercise limitation. If this is physiologically

Figure 1 Section 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 mechanical in origin and should be regarded as a contributory factor to exercise limitation rather than its prime cause. Moreover, breathlessness is the initia-
tor 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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Lung function in preschool children

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

The regression formulæ 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 are younger (fig 3).

The “candle blowing” incentives were as-
sumed to facilitate technically correct spiromet-
tic 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.1,3 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.1 Inclusion of curves with a difference of 10% between the

References


Author’s reply

Experts were given no clinical details except for times of waking and sleeping, and times of starting and leaving work. They were asked to make judgements based on the peak expira-
tory flow (PEF) record alone, similar to the judgements made by the OASYS program. OASYS-2 has been shown to have a sensitivity of around 70% when tested against independ-
ent objective diagnoses (mostly specific bron-
chial provocation testing) and a specificity of 94%. The need is therefore to achieve in-
creased sensitivity.

The experts underscored compared with OASYS-2 and did not appear to be detecting work related changes missed by OASYS-2. In practice, tests are interpreted in the light of clinical information (requiring expertise) but, in our practice, occupational asthma often occurs in unlikely places and is frequently diagnosed when the specific exposures are unknown.

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 is difficult, and non-professionals require expert advice with particular emphasis on record-
ning 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. When the patient returns to clinic with his record stored in an electronic meter. The clin-
ician and patient review the record together. This allows the clinician to ask questions sug-
gested 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 when others do). The clinician is able to interpret the clinical information and record is thus even closer, enhancing the diagnostic toolkit referred to by Dr Fishwick and colleagues.

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Acceptance criteria for correct FVC curves are vague in the absence of expiration time and “end of test” criteria.1 Inclusion of curves with a difference of 10% between the
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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