PostScript

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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 renal angiomyolipomas which are also the most common renal lesion in TSC.[1] LAM occurs in association with TSC or, less commonly, as a sporadic single entity.[2] It is almost exclusive to woman, usually of childbearing age. The most common presentations are dyspnoea, pneumothorax, or chylothorax.[1 2] LAM may be asymptomatic.

A 47 year old woman presented with a 9 month history of haemoptysis on exertion. Dyspnoea was not a feature. Haemoptysis was occurring with increasing regularity following swimming and sexual intercourse, precluding both activities. The volume of blood was usually 10–20 ml. She had stopped smoking a year previously and had a 25 pack year history. There was a history of childhood epilepsy. There was no medical family history.

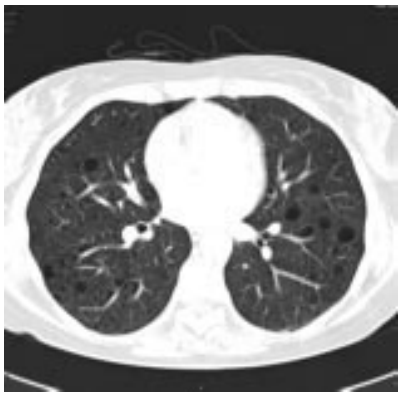

Figure 1 Section from pulmonary high resolution CT scan showing multiple cystic regions with normal intervening lung parenchyma.

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: FEV_1 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 angiomyolipomas.

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.[1] 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*[1] 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 ultrasonography 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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Reference

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Dysfunctional breathing in COPD

I was interested to read Dr Morgan's review of dysfunctional breathing in asthma in the 2002 Year in Review,[1] 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 cardiorespiratory dysfunction is exercise limitation. If this is physiologically

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 initiator 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 associated 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 introduce breathlessness last rather than first when talking to a patient.

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Occupational asthma evaluation

We read with interest the paper by Baldwin *et al*[1] 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 interested 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 mathematical data?

In the clinical setting a decision is made to perform regular PEF monitoring in those patients who are thought to have a reasonable chance of having occupational asthma, as judged by the clinical information to date. Perhaps a further study option would be to give experts the clinical data first (more like the real life situation) and ask for a likelihood of occupational asthma based on this assessment, followed by a revision of that likelihood after PEF data are supplied. Would revealing the work effect score lead to further revision of the perceived estimate? Individual experts may be more or less swayed by the clinical data due to variation in their own practice, types of cases seen, geographical location, and so on.

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 likelihood as well as the PEF pattern. OASYS systems 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.[2] It is therefore important for all such patients in the UK to have access to competent individuals 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 occupational 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.

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 remains 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 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 expiratory 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 independent objective diagnoses (mostly specific bronchial provocation testing) and a specificity of 94%. The need is therefore to achieve increased 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 diagnosis is suspected and before workers are removed from their jobs. Supervising such records does, however, need a degree of expertise with particular emphasis on recording working times, keeping treatment constant, 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 PEF record stored in an electronic meter. The clinician and patient review the record together. This allows the clinician to ask those questions 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 deterioration when others do). The integration of clinical 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*[1] on the feasibility of spirometric tests in preschool children using candle blowing incentives, in support of recent publications.[2–5] As there is a dearth of spirometric 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 symptoms" or "parental smoking habits". It would be interesting to stratify the results, analysing healthy and non-healthy populations separately.

The actual age distribution of the preschool population in table 1 ranged from 4.1 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. Evaluating younger and older children may decrease 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 assumed to facilitate technically correct spirometric tests in the young children. We found that such incentives induced premature termination of forced vital capacity (FVC) which led to lower values than with other methods.[2 3] If this is not the case, how do the authors explain the lower FVC values compared with those of Eigen *et al*,[3] while the forced expiratory volume in 1 second (FEV_1) values were similar (fig 3)?

Acceptance criteria for correct FVC curves are vague in the absence of expiration time and "end of test" criteria.[4 5] 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).[3 6]

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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