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 dermato-logical pathology. Lymphangioleiomyomatosis (LAM) is principally a pulmonary condition characterised by smooth muscle (leiomyo) proliferation around lymphatics (lymph), blood vessels (angi), 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. 2 LAM occurs in association with TSC or, less commonly, as a sporadic single entity. 3 It is almost exclusive to women, usually of child-bearing age. The most common presentations are dyspnoea, pneumothorax, or chylothorax. 4, 5 LAM may be asymptomatic. A 47 year old woman presented with a 9 month history of haemoptysm on exertion. Dyspnoea was not a feature. Haemoptysm 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 other significant medical history.

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. 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 angiomylipomas.

The above findings fulfil the criteria for a diagnosis of LAM and TSC. 6 In view of the diverse clinical course of LAM and the problematic value of hormone therapy, the patient was not commenced on treatment but referred for genetic screening. This case underscores the need to consider such a diagnosis in female patients presenting with solitary exertional haemoptysms.

References

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 vs 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 both in the sitting and supine positions can be very helpful in the diagnosis and follow up of patients with diaphragmatic paralysis.
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, then the mechanism 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 which then to ask about the associated symptoms would lead to a much better approach to the management 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 when talking to a patient.

C K Connolly
Aldbrough House, Aldbrough St John, Richmond, North Yorkshire DL11 7TP; ckk.connolly@medix-uk.com

Reference
1 Morgan MDL Dysfunctional breathing in asthma is it common, identifiable and correctable? Thorax 2002;57(Suppl II):i3-i5–1

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 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 sensitizer 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 risk 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 the perceived estimate? Individual experts the work effect score lead to further revision after PEF data are supplied. Would revealing the clinical information to date, followed by a revision of that likelihood as well as the PEF pattern. OASYS systems clearly involve 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.1 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 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 varies across 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.

D Fishwick, L M Bradshaw, P A Tate, A D Curran
Sheffield Occupational and Environmental Lung Injury Centre, Health and Safety Laboratory, Broad lane, Sheffield S3 7HG, UK; david.fishwick@dh.gov.uk

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 likelihood as well as the PEF pattern. OASYS systems clearly involve 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 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 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 varies across 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.

D Fishwick, L M Bradshaw, P A Tate, A D Curran
Sheffield Occupational and Environmental Lung Injury Centre, Health and Safety Laboratory, Broad lane, Sheffield S3 7HG, UK; david.fishwick@dh.gov.uk

Reference

Lung function in preschool children

We read with great interest the recent paper by Nyström et al 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 4.3 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. 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 asthmatic (fig 1).

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.6–7 If this is not the case, how do the authors explain the lower FVC values compared with those of Egen 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.6–7 Inclusion of curves with a difference of 10% between the

www.thoraxjnl.com

Downloaded from http://thorax.bmj.com/ on July 7, 2017 - Published by group.bmj.com
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.

D Vilozni, O Efrati, A Barak
Sheba Medical Center, Ramat Gan, Israel 52625; avi_vil@netvision.net.il

References

Clinical Evidence—Call for contributors

Clinical Evidence is a regularly updated evidence based journal available worldwide both as a paper version and on the internet. Clinical Evidence needs to recruit a number of new contributors. Contributors are health care professionals or epidemiologists with experience in evidence based medicine and the ability to write in a concise and structured way.

Currently, we are interested in finding contributors with an interest in the following clinical areas:

• Altitude sickness; Autism; Basal cell carcinoma; Breast feeding; Carbon monoxide poisoning; Cervical cancer; Cystic fibrosis; Ectopic pregnancy; Grief/bereavement; Halitosis; Hodgkin’s disease; Infectious mononucleosis (glandular fever); Kidney stones; Malignant melanoma (metastatic); Mesothelioma; Myeloma; Ovarian cyst; Pancreatitis (acute); Pancreatitis (chronic); Polymyalgia rheumatica; Post-partum haemorrhage; Pulmonary embolism; Recurrent miscarriage; Repetitive strain injury; Scoliosis; Seasonal affective disorder; Squint; Systemic lupus erythematosus; Testicular cancer; Varicocele; Viral meningitis; Vitiligo

However, we are always looking for others, so do not let this list discourage you.

Being a contributor involves:
• Appraising the results of literature searches (performed by our Information Specialists) to identify high quality evidence for inclusion in the journal.
• Writing to a highly structured template (about 2000–3000 words), using evidence from selected studies, within 6–8 weeks of receiving the literature search results.
• Working with Clinical Evidence Editors to ensure that the text meets rigorous epidemiological and style standards.
• Updating the text every eight months to incorporate new evidence.
• Expanding the topic to include new questions once every 12–18 months.

If you would like to become a contributor for Clinical Evidence or require more information about what this involves please send your contact details and a copy of your CV, clearly stating the clinical area you are interested in, to Claire Folkes (cfolkes@bmjgroup.com).

Call for peer reviewers

Clinical Evidence also needs to recruit a number of new peer reviewers specifically with an interest in the clinical areas stated above, and also others related to general practice. Peer reviewers are health care professionals or epidemiologists with experience in evidence based medicine. As a peer reviewer you would be asked for your views on the clinical relevance, validity, and accessibility of specific topics within the journal, and their usefulness to the intended audience (international specialists and health care professionals, possibly with limited statistical knowledge). Topics are usually 2000–3000 words in length and we would ask you to review between 2–5 topics per year. The peer review process takes place throughout the year, and our turnaround time for each review is ideally 10–14 days.

If you are interested in becoming a peer reviewer for Clinical Evidence, please complete the peer review questionnaire at www.clinicaleducation.com or contact Claire Folkes (cfolkes@bmjgroup.com).