Exertional haemoptysis: LAM and TSC

Tuberculous 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 dermal angiomyolipomas (LAM) is principally a pulmonary condition characterised by smooth muscle (leiomyoma) proliferation around lymphatics (lymph), blood vessels (angio), and alveolar airways. Cystic destruction of lung parenchyma results in the development of pneumatolysis usually followed by fibrosis. Cardiovascular and respiratory examinations were normal. Pulmonary function tests showed normal lung volumes: FEF 27.2, FVC 3.43, TLC 5.21, and RV 1.96 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.2 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.3 describing the good results of diaphragm plication for diaphragmatic paralysis following blunt trauma. The authors conclude that plication of the diaphragm in the sitting position is helpful in selecting patients for this procedure. They propose that the presence of a chest wall defect and a downward shift are important indicators of phrenic nerve injury.

We would like to point out that plication of the diaphragm in the supine position is a much simpler procedure. We have performed this procedure in a number of patients with phrenic nerve injuries and have found it to be successful in all cases. The technique involves making a small incision in the abdomen and suturing the diaphragm in a horizontal position.

In conclusion, we believe that the technique of diaphragm plication in the supine position is a simple and effective method for the treatment of phrenic nerve injuries.

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References
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 Nystrø et al.1 on the feasibility of spiromet- ric tests in preschool children using candle blowing incentives, in support of recent publica- tions.2–5 As there is a dearth of spiro- metric 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 are asthmatic.

The “candle blowing” incentives were as- sumed to facilitate technically correct spiromet- ric tests in the young children. We found that such incentives induced premature ter- minal deflation of forced vital capacity (FVC) which led to lower values than with other methods.2 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.6 Inclusion of curves with a difference of 10% between the

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 patients does, however, require 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 peak flow record stored in an electronic meter. The cli- nician 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 pattern on the 25th of last month” (when a single work day shows no deteriora- tion 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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two best curves should be avoided on the basis of standard recommendations and previously published data (<5% difference only).\(^1\)

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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