LETTERS

International registry for idiopathic pulmonary fibrosis

We have read with great interest the recent review by Wilson *et al* stating the case for an international registry for idiopathic pulmonary fibrosis (IPF).[1] This timely proposal prompts us to inform the community that, effective from 1 January 2008, the European Commission is funding a new network entitled "European IPF Network: Natural course, Pathomechanisms and Novel Treatment Options in Idiopathic Pulmonary Fibrosis" (*eur*IPF*net*; www.pulmonary-fibrosis.net). Among many other scientific goals, the *eur*IPF*net* will establish a European-wide, internet-based registry (*eur*IPF*reg*) and biobank (*eur*IPF*bank*) for IPF which, in principle, will be open to all interested collegues. The opening of this registry is scheduled for mid 2008 (please follow news on www.pulmonary-fibrosis.net). Key features of this registry will be the collection of all relevant baseline and follow-up clinical data from patients with IPF as well as bronchoalveolar lavage fluid, blood and tissue specimens. Rigorous multidisciplinary verification of the diagnosis will be undertaken by external experts. We plan to quantify the frequency of respiratory infections, extent of pulmonary hypertension, quality of life and response to treatment modalities. Using the collected biomaterials, we intend to investigate novel surrogate parameters of disease progression, establish new disease-specific markers and identify novel candidate genes relevant to the pathophysiology of IPF. Our goals exactly concord with those outlined by Wilson *et al*[1] and we confidently expect that the European IPF registry will foster research on and facilitate the implementation of clinical trials in IPF. Interested collegues are encouraged to participate and preregister (use "contact" on www.pulmonary-fibrosis.net).

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Authors' reply

We are most grateful for the response to our call for an international registry for idiopathic pulmonary fibrosis (IPF) from the European IPF Network collaborators.[1] This most timely registry should be the forerunner of similar disease-specific approaches to problem-solving in health care. The concept of biomaterial collection in conjunction with clinical data should encourage participation from scientists with an ability to contribute to knowledge of the pathogenesis of IPF. We feel this most positive beginning should be a stimulus to other groups, particularly in North America and the Asia-Pacific region, to follow this European initiative.

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REFERENCE

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Strategies to screen for adrenal suppression in children with asthma: there is no consensus among UK centres

Inhaled corticosteroids were designed to avoid the numerous adverse effects of oral corticosteroids in the treatment of asthma. In children, inhaled corticosteroids have been proved to be highly effective and it was initially thought that the risk of adrenal suppression was low.[1] As a result, high "off-licence" doses (eg, ⩾1000 μg/day of fluticasone proprionate) were recommended in difficult cases by national guidelines.[2] The efficacy and safety of such high doses has been seriously questioned by the reporting of around 30 cases of life threatening acute adrenal crisis, including one death, in children maintained on inhaled corticosteroids (largely high dose fluticasone proprionate).[3]

Current guidelines therefore caution that doses ⩾400 μg/day of fluticasone proprionate or equivalent should be prescribed by a specialist who should be aware of the potential for adrenal suppression.[4] A range of tests, varying in invasiveness and complexity, including early morning urinary cortisol, low and high dose synacthen tests and the potentially hazardous insulin–hypoglycaemia test, exist to assess adrenal function in children. It is uncertain, however, which test is most appropriate to detect clinically relevant adrenal suppression in children with asthma.[5] There are other important questions, such as the reproducibility of individual results, threshold doses above which to test, how often to repeat tests or indeed should we test at all?[5 6]

We therefore investigated current practice in screening children with asthma for adrenal suppression in the UK. A postal questionnaire was sent to each of the 23 tertiary paediatric respiratory centres of which 14 responded.

Only eight (57%) centres have an official policy and of these in only 25% is it extended to regional hospitals. In children prescribed fluticasone proprionate, seven (50%) centres test at ⩾500 μg/day, three (21%) at ⩾1000 μg/day and in four (29%) it varies. For beclomethasone, seven (50%) test at ⩾1000 μg/day, two (14%) at ⩾1500 μg/day and ⩾2000 μg/day and it varies in five (36%). Oral prednisolone and nasal sprays were taken into account by eight (57%) and four (29%), respectively. A low dose synacthen test is performed by seven (50%), three (21%) high dose synacthen test, one (8%) morning cortisol and in three (21%) it varies. Five different abnormal cortisol responses are used. Tests are repeated annually by 10 (71%), two (15%) test 6 monthly and only once, respectively. Steroid cards are issued by eight (57%) of the centres. In total eight (57%) of the respondents regarded adrenal suppression as a significant problem and nine (64%) have changed their practice over the past 5 years.

We therefore conclude that there is no national consensus in the UK on screening of children with asthma for adrenal suppression. Specific areas of divergence include: the threshold dose to start testing, which test to perform, how to interpret the results and when it should be repeated. Further studies and discussions are required to establish an evidence base about how best to screen for this potentially life threatening problem.

Irrespective of the screening policies used in different centres, it is vital that the assumption is made that a child may be adrenally suppressed unless there is clear evidence that this is not the case. Issuing of steroid information cards to children and families is one method of reinforcing this fact. Only just over half of responding centres pursue such a policy. It should be noted however that there is little published evidence to show benefit, or equally detriment, from the use of steroid cards.

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