

Selected abstracts from Paediatrics: things you don't grow out of (managing chronic disorders)

CHRONIC RESPIRATORY DISEASE

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Background The chronic respiratory problems that we pass on to our adult colleagues vary from asthma, which is becoming so common that we will soon have to be arranging counselling for non-asthmatics, to a number of much less prevalent syndromes such as ciliary motility disorders and sarcoidosis.

In the middle of this prevalence range is cystic fibrosis, whose management is an excellent exemplar of the approach to many chronic disorders, comprising antenatal and postnatal screening and diagnosis, breaking the bad news, encouraging parental involvement in management and in management decisions, deploying scarce resources as usefully as possible, providing long-term support for patient and family, handling all manner of medical and psychosocial crises and finally providing sensitive and appropriate terminal care.

Methods or theme In common with other chronic disorders, respiratory disorders give rise to a broad range of problems in addition to those related to medical management. Parental fear, guilt and shame attend the child's negative self-image, feelings of isolation and loneliness, and sometimes overt psychiatric disorder. However, although these problems are common to all chronic disease processes, they reflect the complexities inherent to each specific disease, and supportive management requires not only psychological diagnostic and counselling skills but also a detailed knowledge of the disorders themselves.

There is therefore a need not only for skilled counselling and other supportive services to be provided for chronic respiratory disorders, but also for such support to be given by individuals who are knowledgeable about disorders themselves.

In asthma, perhaps the most common problem is impaired quality of life resulting from poor compliance with therapy. This in turn may result from embarrassment about using inhalers in public, or even criticism from others about drug usage. Psychological problems are common, and were probably better managed when asthma was considered a psychosomatic disorder.

In cystic fibrosis, there are major problems with self-esteem and self-image, with the imposition of therapeutic

regimens of great intensity and complexity, with issues surrounding infertility and child-bearing and with issues related to mortality.

Conclusion These conditions should be seen not as simple medical problems to be managed pharmacologically, but as complex multidisciplinary problems.

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Keywords: Asthma, cystic fibrosis, complications, counselling, psychosocial problems, psychosomatic disorder

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PAEDIATRIC NEUROLOGY

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Background Established neurological disorders of childhood stay with you for life. There are few chronic neurological disorders that can yet be cured, although progression can be modified and symptoms ameliorated. Survival and life expectancy have improved, and that in itself brings new problems of care and provision of services into young adult life.

There are transient neurological disorders in childhood that disappear spontaneously with time. Examples include global developmental delay at the slow end of the normal spectrum, and benign epilepsies of infancy, which affect a genetically predisposed, immature brain.

Treatment for certain brain tumours now promises cure, but there are still the risks associated with radiotherapy and chemotherapy.

Prevention of acquired brain damage in children has had some success, for instance haemophilus and meningococcal immunisation and seat-belt laws, but there are still too many pedestrian head injuries in the UK and non-accidental brain injuries.

I discuss three common childhood conditions, two of which are lifelong – cerebral palsy and Duchenne Muscular Dystrophy (DMD). Survival in severe cerebral palsy and DMD is improving, and new cohorts of children are graduating into adult services. This makes

greater demands on health and social support, and introduces ethical issues about getting the balance correct between survival and quality of life.

The third condition, epilepsy, may change and improve as the child goes through adolescence, but still has the potential to affect quality of life in adulthood.

Keywords: Brain tumours, cerebral palsy, Duchenne Muscular Dystrophy, epilepsy, neurological disorders, pedestrian head injuries, transient neurological disorders

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Declaration of interest: None declared.

MAKING DIFFICULT DECISIONS: USING AN ETHICAL FRAMEWORK

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Background Ethical dilemmas arise when clinical or other facts cannot determine what should be done, when moral obligations conflict or when the law is ambivalent or silent. They may range in scope from complex end-of-life issues to simple matters of truth-telling over practical procedures, e.g. venepuncture.

The function of ethics is to define and analyse dilemmas, and apply moral theories or principles in an attempt to assist their resolution. There may be more than one or no 'right' answers.

Traditional moral theories include those in which an action's consequences determine its rightness and those

where action is determined by rational consideration of duties. More frequently an analytical approach involving the four *prima facie* principles of beneficence, non-maleficence, respect for autonomy (the right of self-determination) and fairness is used. It may be difficult to determine which principle should be paramount. Good decision-making is characterised by inclusiveness, transparency, reasonableness and accountability.

From the above we can derive two principles that underpin medical practice:

- Act in the best interests of patients
- Obtain valid consent for all interventions

A framework for approaching moral dilemmas includes:

- determining the relevant clinical and social facts
- deciding an appropriate decision-making process
- ascertaining what the law/professional guidance says
- identifying the ethical issues and morally relevant principles
- listing the options and identifying the moral arguments for and against
- choosing an appropriate option and testing whether it can be rebuffed

Outcomes should be reviewed. The experience gained from ethical 'debriefing' may be helpful.

Keywords Accountability, analyse dilemmas, beneficence, conflict, ethical dilemmas, fairness, inclusiveness, law, moral obligations, non-maleficence, reasonableness, respect for autonomy (the right of self-determination), transparency

Sponsorship: None.

Declaration of interest: None declared.

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Selected abstracts from: Respiratory Medicine Symposium 2008

HOW PATIENTS POISON THEIR LUNGS

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Background The last century witnessed the ascent of cigarette smoking as the major form of tobacco consumption and, as a consequence, the rise in smoking-related diseases. Smoking already kills more than one in ten adults worldwide and by 2030 it will kill an estimated 10 million, accounting for one in six individuals. Much of the work undertaken by respiratory physicians deals with the impact of tobacco smoking. In particular, the majority of cases of lung cancer and chronic obstructive pulmonary disease (COPD) are directly attributable to the habit; however, smoking is also an important contributor to an increased risk of respiratory infections and inadequate control of asthma. Although there is some optimism that cigarette consumption is declining, alternative forms of smoking (which are likely to be more dangerous) are becoming more popular, such as bidis and kreteks.

Many illegal drugs are also smoked and delivered directly to the lung. Cannabis is the most commonly used drug in Scotland, particularly among the young. Several papers have linked cannabis smoking to the development of respiratory symptoms, impaired lung function, the appearance of bullous emphysema and an increased risk of respiratory infections. In addition, there are concerns that cannabis smoking may be accompanied by an increased risk of lung cancer. Crack is becoming an increasingly popular way of taking cocaine and may be an important contributor to the presentation of asthma, as may also be the case with heroin. When taken by the intravenous route these drugs can give rise to talcosis and septic emboli.

Further reading

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Keywords Bidis, cannabis, cocaine, heroin, tobacco

Sponsorship None.

Declaration of interest None declared.

HOW EMPLOYERS POISON THE LUNGS OF EMPLOYEES

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Background Occupational lung disease is common and a preventable cause of respiratory disability. While in the UK some forms of the disease are declining with loss of the relevant industry (e.g. coal mining), others are becoming more apparent (e.g. the possible contribution of chemicals to occupational COPD).

Methods or theme Occupational asthma shows little sign of reducing in prevalence according to the Surveillance of Work-Related and Occupational Respiratory Disease (SWORD) data for the UK, while the costs of the disease to UK taxpayers and the patients themselves are high. Recognition of the occupational contribution to asthma and COPD in primary care is generally poor, and approaches to raise awareness in the community have the potential to identify cases early and reduce morbidity. The advent of the European Union Registration, Evaluation and Authorisation of Chemicals (REACH) legislation may have implications in Europe in assessing to what extent occupational and non-occupational chemicals might contribute to occupational airways disease. However, with around 15% of the burden of COPD being attributed to occupation, steps to recognise and reduce relevant exposures would have important benefits in terms of costs to the NHS and quality of life for those affected.

Conclusions Early identification in primary care of occupational airways disease and the involvement of employers in recognising and reducing risks would have an important impact on the morbidity from occupational airways disease.

Further reading

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Keywords Occupational asthma, occupational COPD, reactive airway dysfunction (RADs)

Sponsorship None.

Declaration of interest Professor Ayres has no conflicts of interest save that he undertakes medico-legal work for both plaintiffs and defendants in cases of occupational lung disease.

HOW DOCTORS POISON THEIR PATIENTS' LUNGS

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Background Lung injury as a consequence of prescribed medication has a long and notorious history. While drugs can cause, or exacerbate, all recognised patterns of lung dysfunction, it is the capacity to induce histopathological patterns of interstitial pneumonia that is of particular interest. By careful dissection of the mechanisms by which drugs and other therapies lead to alveolar epithelial injury, lung inflammation and aberrant repair, it may be possible to gain crucial insights into the pathogenesis of the 'idiopathic' interstitial pneumonias, including idiopathic pulmonary fibrosis. For many drugs in clinical use for decades, such as amiodarone and methotrexate, both the mode of action and the mechanism by which lung injury is created are poorly understood.

Recent experience with a new generation of therapies directed towards defined molecular targets, for example epidermal growth factor receptor (EGFR) and tumour necrosis factor- α (TNF- α), has shown that they too are associated with lung toxicity. Since small molecule and antibody-based agents have been designed to be highly target-specific, the unintended biological effects in humans serve to illuminate the true importance of specific cell-signalling pathways that hitherto have been debated only on the basis of in-vitro studies or animal models. Perhaps most remarkable is the observation that, despite interfering with pathways that are often considered critical to maintaining homeostasis, serious toxicity is quite uncommon. The increasing use of these therapies, however, means that chest physicians must feel confident in the diagnosis and management of patients with occasionally catastrophic 'biological' drug-induced lung disease.

Finally, 'nano-medicine' is a rapidly emerging prospect for the future. Early studies offer the hope of improving the safety profile of a generation of drugs of proven efficacy, but as yet so little is known of the biology of complex nanosystems that the hype should be countered by the potential for causing serious harm.

Further reading

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Keywords Biological therapy, drug-induced lung disease, interstitial pneumonia, nano-medicine

Sponsorship None.

Declaration of interest None declared.

CHANGES IN THE BURDEN OF RESPIRATORY DISEASES UP TO 2050

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Background Although it is difficult to extrapolate into the future, trends in disease can be broken down into effects that occur early in life and persist (either because they have a permanent effect or because the exposure itself tends to persist), effects that occur close to the time of the exposure and are quickly reversible, and effects that are due to ageing. This form of analysis provides a way of identifying some trends that are likely to persist into the medium term at least. This method will be illustrated from studies of atopy and asthma, COPD and mesothelioma.

Methods or theme The prevalence of asthma and wheezy illness has increased rapidly over the last half century as measured by symptom questionnaires. More recently, surveys among children have shown some levelling off or decrease in prevalence in the UK. More significantly, however, the prevalence of atopy can be shown to have increased rapidly from generation to generation, from those born in the 1920s until at least the 1970s. Whatever happens among children, the prevalence of atopic disease among adults is likely to continue to increase for some time to come.

In the UK, age-adjusted rates of COPD mortality are falling. The worldwide prediction, however, for numbers of deaths and proportions of deaths is that these will increase. There are two reasons for this prediction. The first is the great increase in the smoking of manufactured cigarettes in countries with low levels of average income, and the second is the epidemiological and demographic transitions of the population structures of these countries to ones with low infant mortality and older populations.

In the case of mesothelioma, the predictions are that the total number of deaths will continue to rise for a while, but the age-adjusted mortality in lower age has been falling for some time. This coincides with the change in exposure to asbestos, with import restrictions starting in the 1970s.

Conclusions Although predictions can be upset by events, there are some trends that are likely to be very difficult to change in the short term.

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Keywords Asthma, atopy, COPD, mesothelioma, trends
Sponsorship None.

Declaration of interest Professor Burney has received lecture fees from Astra-Zeneca over the last two years.

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