30 Paediatric respiratory medicine

Introduction

Key points

- Paediatric respiratory medicine (PRM) was only recognised in some EU countries as a subspecialty of paediatrics in the 1970s; in many countries, there is still no such specialty.
- In small hospitals, all paediatricians care for children with acute respiratory illness. Only large city hospitals have a dedicated paediatric respiratory medicine team, and few offer expertise in highly specialised areas.
- Training in paediatric respiratory medicine varies greatly between countries – from an established course of several years to a simple determination of competence by a supervisor, regardless of period of training. The Paediatric HERMES syllabus aims to harmonise training standards across Europe.
- The investigation and management of several conditions has now been standardised, but there is no internationally agreed guideline for many conditions, and those guidelines that do exist are often based on consensus rather than evidence.
- Across Europe, there are obvious disparities in the number of PRM specialists and in the levels of morbidity and mortality from childhood respiratory conditions; these disparities should be addressed.

Paediatric respiratory medicine (PRM) is a multidisciplinary subspecialty within the specialty of paediatrics, involving doctors, nurses, respiratory physiologists, physiotherapists, the child and the parent/caregiver. PRM first emerged as a subspecialty in some European countries in the 1970s when the European Paediatric Respiratory Society (EPRS) was formed. The EPRS and the Paediatric Assembly of the European Respiratory Society (ERS) existed alongside each other in the early 1990s, until the EPRS was incorporated into the ERS Paediatric Assembly in 1993. The major reason PRM emerged relatively late as a subspecialty of paediatrics was because respiratory problems were so common in children that all paediatricians were expected to be specialists in their diagnosis and management. While PRM is an established subspecialty in some European Union (EU) countries, this is by no means the case in all countries; countries that do not recognise PRM as a subspecialty include Finland, Greece, Italy and Spain. Among the EU countries in which PRM is recognised as a subspecialty, the typical ratio of paediatric to adult respiratory physicians is approximately 1:10, the same as the ratio of children to adults in the population. However, in some countries, the ratio of paediatric to adult respiratory physicians is as low as 1:50 (figure 1). The number of PRM specialists is not known in some countries, and it is likely that in many, there are simply no such specialists.

The aim of this chapter is to give an overview of PRM across Europe and not to focus on specific conditions.

> *Antibiotic doses are based on studies in adults with the assumption that children are small adults […] children therefore often receive inadequate doses of medication*

As with all medical areas, PRM is in a state of constant evolution due to several different drivers (summarised in table 1). The present chapter therefore captures the situation in Europe in 2013 and is broken down into sections considering setting, training, standardisation of care, and developments, and ends with a brief summary that includes recommendations for the future.

Setting

Children are usually defined as individuals who are ⩽16 years of age. This age group experiences frequent respiratory symptoms: all children will cough (mostly due to respiratory infections) and up to one half have had wheezing by 5 years of age. Children, in particular those under 5 years of age, have the highest burden of respiratory symptoms of all age groups in the general population. The majority of paediatric respiratory illnesses are mild and resolve on their own or respond to treatment given in primary care. In most small hospitals, all paediatricians care for children with acute respiratory illnesses, although some paediatricians may have a special interest in respiratory paediatrics and work with a PRM team in their regional centre. A small proportion of children with severe and/or persistent respiratory problems will be cared for by a regional PRM team. Referral to the PRM team can come from a general practitioner or a hospital doctor who does not specialise in PRM; in some countries, parents can arrange an appointment with the PRM team directly.

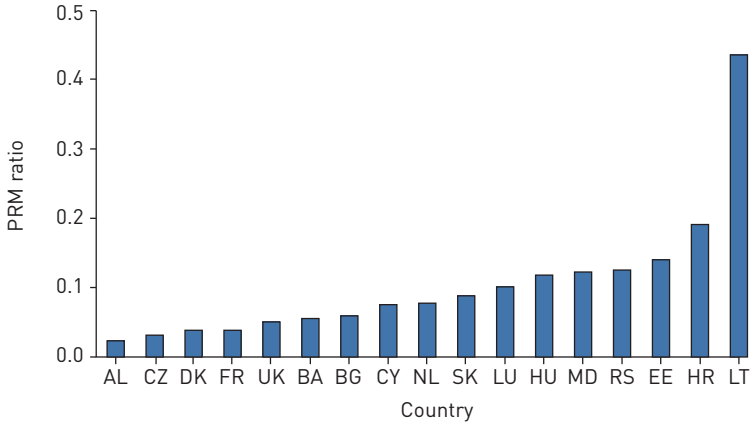

Figure 1 – The ratio of specialists in paediatric respiratory medicine (PRM) to specialists in adult respiratory medicine. In addition to the 17 countries shown, no data were provided for 24 countries and in seven, PRM is not recognised as a subspecialty.

Driver	Example
Changing epidemiology	Asthma 'epidemic' during 1980s and 1990s
Changing expectations	Expertise in paediatric respiratory medicine cannot always be provided by general paediatricians
New treatments/interventions	Management of sleep breathing disorders
Changes in working patterns	EU working time directive

Table 1 - Changes that have an impact on paediatric respiratory medicine. EU: European Union.

"Paediatric respiratory medicine is not recognised as a subspecialty in many European countries, including Finland, Greece, Italy and Spain"

Almost all PRM teams are based in large city hospitals; figure 2 indicates where interactions with other specialties may occur. Expertise within the respiratory team includes the following areas: asthma, cystic fibrosis, sleep breathing disorders, intensive care, noninvasive ventilation, bronchoscopy (a flexible telescope used to examine the airways) and research. Individual members within a single PRM team will have expertise in several areas and, depending on the number of individuals within the team, all areas may be covered in one hospital; in some hospitals, certain areas of expertise, such as bronchoscopy and sleep breathing disorders, may not be covered. Very few hospitals demonstrate expertise in certain highly specialised areas; caring for children before and after lung transplantation, for example. Clinical areas such as allergy, neonatology, paediatric intensive care and infectious disease have considerable overlap with PRM but are subspecialties in their own right in many countries (although allergy and PRM are considered a single specialty in some countries).

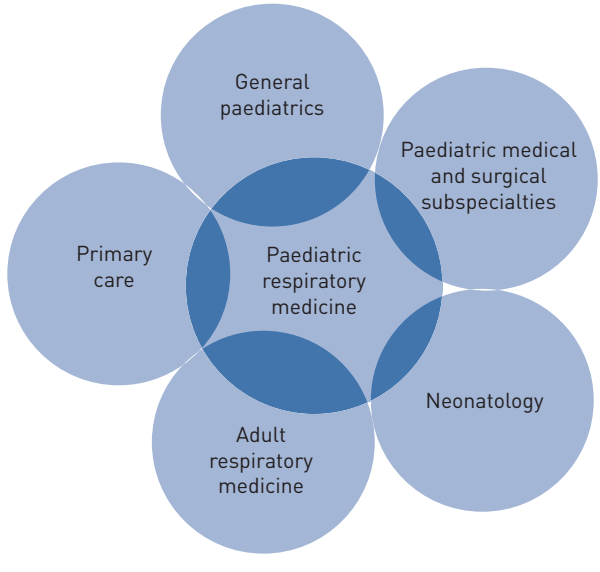

Figure 2 – Interactions between paediatric respiratory medicine and other medical specialties.

Training

When is a trainee qualified to be a specialist? This question was put to paediatric respiratory groups across Europe approximately 10 years ago and perhaps unsurprisingly, the answers were variable. Many European countries were only able to give a very rough estimation of the length of the period of training – ranging from 'a few' to 'several' years. Some smaller countries had no formal training and trainees often went overseas for experience, while in at least one country competence was determined by the supervisor regardless of the period in training. Training in different countries will always reflect national requirements and some differences are inevitable; however, EU legislation means that qualifications in different countries are recognised as equal and therefore consistency in training across the EU is highly desirable. In an attempt to move towards harmonised training, the ERS developed a common syllabus for PRM trainees between 2002 and 2009 – the Paediatric Harmonised Education in Respiratory Medicine for European Specialists (HERMES; hermes.ersnet.org). This included an examination, which applicants first sat in 2011.

Before beginning training in PRM, trainees are expected to have a minimum 3 years' experience in general paediatrics. Training in PRM usually lasts a further 3 years. Many trainees also complete an additional 2 or 3 years of training in research. At the time of writing, the Paediatric HERMES syllabus and examination were not compulsory but, in future, trainees who have passed the examination are likely to be seen as 'stronger' candidates when applying for specialist positions. The content of the Paediatric HERMES syllabus includes 21 mandatory modules and three optional ones (table 2).

Standardisation of care: towards a common goal

In addition to training, the management of specific conditions is also being standardised throughout Europe and across the world. The need for standardisation of management was illustrated by a paper published in 1998, which compared the management of a common respiratory infection in infancy (bronchiolitis) in many European countries and in the USA. The main factor linked to the length of stay in hospital was not severity of illness but the country in which the infant lived. A 2010 study of cystic fibrosis across Europe identified fewer individuals surviving to adulthood in non-EU compared with EU countries, and suggested that up to 50% of all cases of cystic fibrosis in non-EU countries were not being diagnosed (and by implication, not receiving appropriate treatment). The considerable variation between countries, and the desire to standardise PRM training, has driven the standardisation of treatment and investigation. Table 3 lists some of the areas in which guidelines have been established.

The investigation and management of several conditions has now been standardised. However, it is important to note that for most conditions, no internationally agreed guideline exists, and many of the guidelines that do exist are based on consensus (*i.e.* what is currently being done) and rarely on an evidence base (*i.e.* clinical trials). To take drug dose as an example, most antibiotic doses are based on studies performed in adults with the assumption that children are small adults; however, this is often not the case and generally children receive inadequate doses of medication. There is a pressing need for clinical trials in PRM and this has been recognised by the 2006 EU Clinical Trials Directive, which obliges the pharmaceutical industry to test new medications in children as well as adults.

In some countries, the ratio of paediatric to adult respiratory physicians is as low as 1:50

Mandatory	
	Respiratory symptoms and signs
	Pulmonary function testing
	Airway endoscopy
	Imaging
	Acute and chronic lung infection
	Tuberculosis
	Cystic fibrosis
	Bronchial asthma
	Allergic disorders
	Congenital malformations
	Bronchopulmonary dysplasia
	Rare diseases
	Sleep medicine
	Rehabilitation
	Inhalation therapy
	Technology-dependent children
	Epidemiology and environmental health
	Management and leadership
	Teaching
	Communication
	Research
Optional	
	Rigid and interventional endoscopy
	Post lung transplant management
	Additional diagnostic tests

Table 2 - Modules of the Paediatric HERMES syllabus. HERMES: Harmonised Education in Respiratory Medicine for European Specialists.

Keeping up with developments

Changes in the conditions that affect children, new therapies and societal expectations mean that PRM is constantly moving forward. Table 4 presents examples of the way in which changing attitudes to paediatric conditions have revolutionised treatment and improved survival for many. Ensuring that children with life-limiting conditions, such as cystic fibrosis and muscular dystrophy (a condition causing weakness of the muscles, including the muscles of breathing), survive into adulthood is one of the successes of PRM, and liaison

Condition	Society/body responsible	Guidelines
Asthma	BTS and SIGN	BTS and SIGN: British Guideline on the Management of Asthma: a National Clinical Guideline
	GINA	International guidelines: www.ginasthma.org/
	iCAALL	International consensus on (ICON) pediatric asthma
	PRACTALL	Diagnosis and treatment of asthma in childhood: a PRACTALL consensus report
Bronchiolitis	SIGN	Bronchiolitis in Children: a National Clinical Guideline
	American Academy of Pediatrics	Diagnosis and management of bronchiolitis
Cystic fibrosis	Cystic Fibrosis Trust	UK guidelines: www.cftrust.org.uk/aboutcf/publications/consensusdoc/
	European Cystic Fibrosis Society	European consensus statements: www.ecfs.eu/publications/consensus_reports
	Cystic Fibrosis Foundation	US guidelines: www.cff.org/treatments/CFCareGuidelines/
Spirometry in preschool children	ATS/ERS	An official American Thoracic Society/European Respiratory Society Statement: Pulmonary function testing in preschool children

Table 3 – Guidelines for diagnosis, testing and management of paediatric respiratory conditions. The list is not exhaustive. Guidelines in Europe and the USA are very similar. BTS: British Thoracic Society; SIGN: Scottish Intercollegiate Guidelines Network; GINA: Global Initiative for Asthma; iCAALL: International Collaboration in Asthma, Allergy and Immunology; PRACTALL: Practicing Allergology or Practical Allergy; ATS: American Thoracic Society; ERS: European Respiratory Society.

with colleagues in adult respiratory medicine has been important and successful. One emerging subspecialty in adult cardiac medicine is care for adults born with congenital heart conditions; in future, there is likely to be more demand for adult respiratory physicians with expertise in conditions such as cystic fibrosis and bronchopulmonary dysplasia (a condition in infants born very prematurely and caused partly as a side-effect of being on a ventilator and partly due to the lungs having to develop outside the womb). In future, a better understanding of the treatment of rare but serious conditions in PRM (often termed 'orphan lung diseases'), such as bronchiolitis obliterans, will emerge as colleagues in PRM across Europe collaborate.

A final aspiration is that in the future, children with and without respiratory problems will be able to breathe better-quality indoor and outdoor air. It is hoped that this will be achieved through legislation aimed at reducing children's exposure to second-hand smoke and car exhaust fumes. The poet William Wordsworth observed that 'The child is the father of the man' and we know that the origins of many adult chest conditions, including asthma and chronic obstructive pulmonary disease, are determined in early life. Improving the quality of air entering children's lungs every time they breathe has to be a priority.

> *There is a pressing need for clinical trials in paediatric respiratory medicine*

Condition	Incidence	Approximate births in Europe n	Previous expectations	Current expectations	Implications
Cystic fibrosis	1 per 3000 births	1700 per year#	1950s: death in pre-school years was usual	Half of babies born in 2012 can expect to live beyond 50 years of age	There are more adult patients with CF than children
Bronchopulmonary dysplasia	1 per 3000 births	1700 per year#,¶	1970s: nothing was done to help babies born at ⩽28 weeks with breathing difficulties	90–95% likelihood of survival if born at 28 weeks; the majority survive without neurological handicap. Risk of neurological problems increases with greater prematurity.	Implications of bronchopulmonary dysplasia for adult life unknown. More children with neurological problems become adults.
Muscular dystrophy	1 per 4000 males	1250 per year#	1990s: death in teens due to respiratory failure	Survival into 20s with noninvasive ventilation	Death from associated heart defects

Table 4 – Conditions in which expectations have changed with implications for paediatric and adult respiratory medicine. #: assuming 5 million deliveries per year in Europe. ¶: assuming 1 per 1000 born at ⩽28 weeks' gestation and one-third of these have bronchopulmonary dysplasia.

Conclusion

PRM is a busy subspecialty that is important to the work of many other clinical groups. Across Europe, there remain obvious disparities in the number of PRM experts and in the levels of morbidity and mortality from childhood respiratory conditions; these disparities should be addressed. Looking ahead, in the coming years, the PRM community aims to continue to work with patient groups and funders to deliver clinical trials upon which to base best practice of care and in order to lobby for improvements in air quality. As with all things, this will require time, money, and most of all, leadership.

Future recommendations

- Greater recognition of PRM as a subspecialty across Europe. There should be PRM specialists in every country as well as training in PRM.
- Narrowing of the gap between the highest and lowest national burden of respiratory morbidity and mortality across Europe, including standardisation of diagnosis and treatment.
- Acknowledgement of the need for researchers to design and funders to support clinical trials in PRM for pharmacological and nonpharmacological interventions.
- Greater recognition of the effect of early exposure on lifelong respiratory wellbeing, including improvement of indoor and outdoor air quality and prevention of smoking in children in Europe (and the rest of the world).

Further reading

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