

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.

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

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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.

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

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).

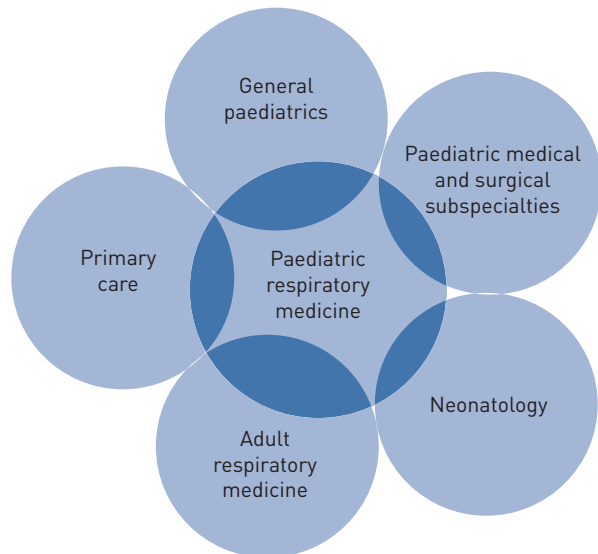


Figure 2 – Interactions between paediatric respiratory medicine and other medical specialties.

Training

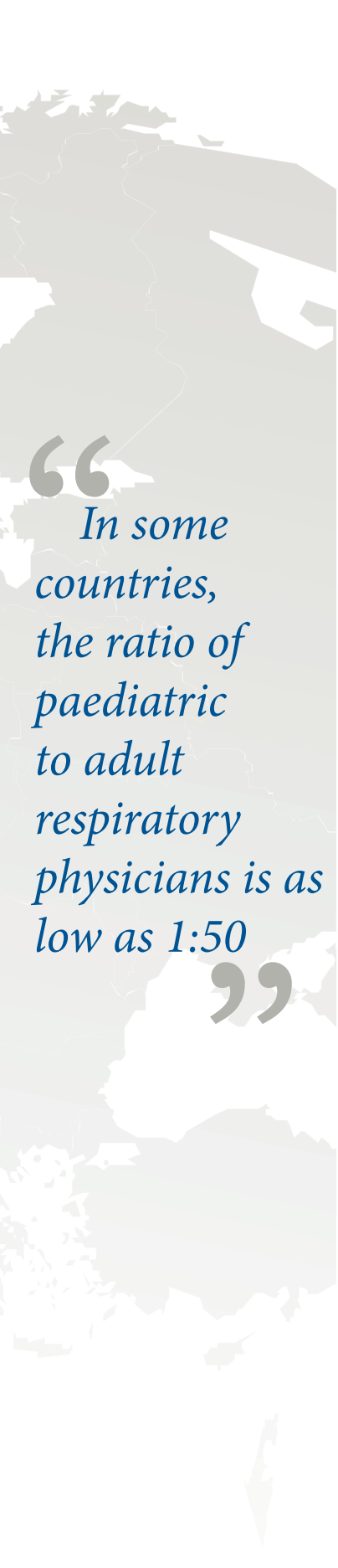
When is a trainee qualified to be a specialist? This question was put to paediatric respiratory groups across Europe approximately 10 years ago and perhaps unsurprisingly, the answers were variable. Many European countries were only able to give a very rough estimation of the length of the period of 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.



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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: Practising 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.

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There is a pressing need for clinical trials in paediatric respiratory medicine
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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 ^{#,1}	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. 1: 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).

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Respiratory intensive care

Introduction



Key points

- Intensive care medicine has progressed considerably and improved survival after long-term ventilation has led to a greatly increased demand for intensive care facilities.
- Historically, intensive care units were led by anaesthetists. Increasingly, however, respiratory intensive care is supervised by respiratory physicians.
- Respiratory intermediate care units and ventilator-weaning units are likely to grow in size and number as the number of patients surviving prolonged ventilation increases and demands on conventional intensive care units grow.
- The technology of extracorporeal oxygenation has improved considerably and the development of an artificial lung has become a realistic prospect.

In recent decades, no branch of medicine has made more progress than intensive care. Important advances include: technological developments to overcome single-organ failure (for example, the development of respirators and extracorporeal oxygenation to treat acute lung failure); improvements in pharmacotherapy based on better understanding of the underlying pathophysiology; and new or improved monitoring systems for surveillance of organ function and to help to direct therapies.

As a consequence of these technological, diagnostic and therapeutic advances, demand for intensive care medicine has escalated continuously. Increasingly, elderly patients with multiple comorbidities are now treated in intensive care units and the boundaries of possible treatments are ever-widening. As recently as 30 years ago, patients needing invasive ventilation for more than 3 days were unlikely to survive. Today, patients requiring long-term ventilation are a common occurrence in every intensive care unit. With the expansion of mechanical ventilation and other organ-replacement techniques, large new areas of medicine have been added to classical acute intensive care. The differences between the 'high-tech' intensive care unit and regular wards have widened. Intermediate care units have also become established, with better monitoring and a greater therapeutic spectrum than is found in conventional wards, but without the organ-replacement techniques of the intensive care unit.

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Respiratory physicians in many countries have established, or are now involved in, specialised weaning centres

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Weaning from prolonged mechanical ventilation is an important aspect of modern intensive care medicine. Specialised units have grown up in many hospitals (long-term care facilities/weaning units), and home mechanical ventilation has also expanded greatly in recent years.


New professions have developed, such as the respiratory therapist. However, the development of legal and administrative structures associated with intensive care medicine, the skills of the staff and the definitions of their job profiles have not always kept pace with the rapid changes in medical reality. Historically, the structure of intensive care medicine developed differently in different European countries. Consequently, there are variations in professional responsibility, recruitment and training of staff, career perspectives in intensive care medicine and quality standards.

With the growth of intensive care units, intensive care medicine has become economically more important for hospitals and, consequently, for the financing of the whole healthcare sector. These developments will continue, increasing the pressure to create adequate legal and administrative structures in this area. At the same time, the need for qualified specialised personnel and for internationally equivalent and mutually recognised training programmes will increase.

Standards of end-of-life care and decision-making have become an increasingly important issue in intensive care medicine, in particular in patients with end-stage respiratory failure. A European Respiratory Society task force found that in European respiratory intermediate care units and high-dependency units this is highly relevant: an end-of-life decision was taken in 21.5 % of the patients admitted.

The role of respiratory medicine in intensive care

Modern intensive care medicine began in the 1920s with the introduction of the iron lung for the treatment of respiratory failure associated with polio. This expands the lungs with each breath by applying a suction (negative) pressure around the trunk. The next important step was the availability of artificial airway tubes and positive-pressure ventilators, which deliver air directly into the patient's airways. These developed from the requirements of modern surgery to facilitate better control of anaesthesia during operations. Intensive care medicine benefitted from the new ventilation techniques, which at first were applied exclusively to surgical patients.



By the mid 20th century intensive care medicine consisted primarily of mechanical ventilation *via* an endotracheal or tracheostomy tube. In the USA, intensive care became an integral part of respiratory medicine but in most European countries, anaesthetists (anaesthesiologists) became responsible for most general medical and postoperative intensive care units. However, this pattern did not develop uniformly across Europe. In some countries a new specialty of intensive care medicine developed, while in some others training in intensive care was incorporated in the curriculum of several specialties, including anaesthetics (anaesthesiology), internal medicine, surgery and paediatrics. Within the medical specialties more specialised care facilities were developed to care for patients with failure of a single bodily system, *e.g.* coronary care, respiratory care and, latterly, stroke units. In some countries, the representation of respiratory medicine in intensive care is still relatively low, even though acute respiratory failure is one of the three main issues in modern intensive care medicine, along with circulatory failure and severe infection.

The increasing focus of respiratory medicine on patients with chronic respiratory disease has led to the development of respiratory intermediate care units, which specialise in treating respiratory insufficiency as a single-organ problem. Therapeutic techniques were developed that could be performed outside fully equipped intensive care units, including specialised physiotherapy, respiratory therapy, mechanical support systems for the expectoration of secretions, and improved oxygen delivery systems.

The introduction of noninvasive ventilation (NIV) about 20 years ago was a major advance. A face-mask or other device is used to deliver air without the need for intubation and its attendant risks such as infection. Initially, NIV was applied to patients with chronic ventilatory insufficiency caused by diseases that do not originate from the lung, *i.e.* neuromuscular disorders such as the postpolio syndrome and muscular dystrophies, and severe deformation of the thorax and vertebral column such as scoliosis (figure 1). In these patients, ventilation is inadequate due to failure of the 'ventilatory pump', which results in hypercapnia (high arterial carbon dioxide pressure); this is readily corrected by use of NIV applied intermittently (usually during sleep), resulting in a tremendous improvement of quality of life and survival. Long-term domiciliary NIV is now used routinely to treat these conditions.

With better understanding of the pathophysiology of severe chronic obstructive pulmonary disease (COPD) (which results in hypercapnia due, in part, to relative weakness of the respiratory muscles), the use of NIV has been extended

to patients with advanced COPD and hypercapnic respiratory failure, particularly during acute exacerbations (AE-COPD). These patients are a large group, and in many hospitals NIV is now used routinely to treat patients with hypercapnic, acidotic AE-COPD outside traditional intensive care units, in high-dependency units or in the general respiratory ward setting. For many such patients, this avoids the need for intubation and its complications, in particular infection and difficulty in weaning from the ventilator. Long-term home NIV is also used in some patients with COPD and chronic hypercapnia (figure 1), but the optimal indications and selection criteria are the subject of ongoing research. In intensive care units NIV is also often used during tracheal intubation prior to commencing full assisted ventilation, during weaning from mechanical ventilation and after failure of extubation.

Increasing numbers of elderly patients undergo major surgery or need intensive care because of comorbidities or for other reasons. Improvements in ventilation strategies and the consequent reduction of ventilator-associated lung injury, in combination with advances in intensive care medicine, have allowed much longer periods of ventilation. In the past, unless patients were extubated after 3 days they usually died, but now intubation and ventilation for weeks or months is feasible. However, weaning from mechanical ventilation becomes more difficult with every additional day of ventilation. During prolonged mechanical ventilation, patients experience progressive wasting of skeletal muscles and may also develop a peripheral neuropathy, both of which can affect the muscles of the ventilatory pump. Respiratory physicians in many countries have established, or are now involved in, specialised weaning centres. Relevant teaching courses have been developed for physicians, nurses, physiotherapists and respiratory therapists.

Future prospects

Intensive care medicine is likely to be responsible for an increasing proportion of the care of hospitalised patients in nearly all medical areas in the next few years. It will be a major, if not the largest, economic factor for all hospitals. It is likely that intensive care medicine will become even more specialised (figure 2). In addition to the classical

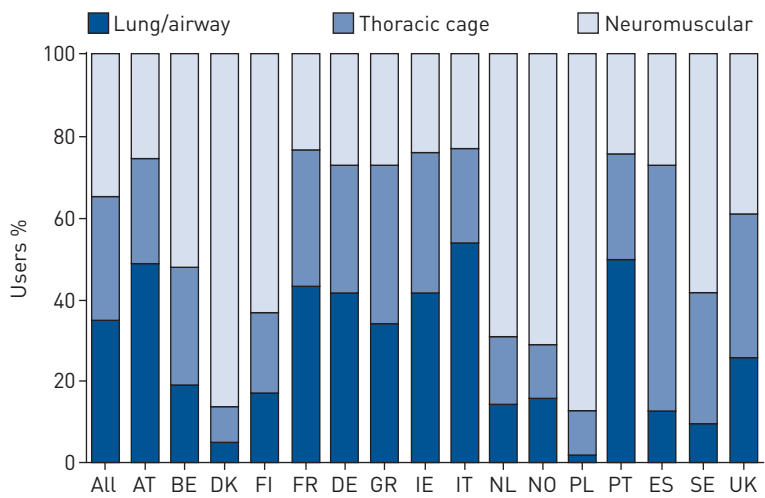


Figure 1 – Indications for home mechanical ventilation in Europe. Reproduced from LLOYD-OWEN *et al.*, 2005.

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The overall relationship between respiratory medicine and intensive care medicine across Europe is unclear
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management of acute and life-threatening complex problems in conventional intensive care units, with high staffing ratios and modern technical equipment, a considerable increase is likely in the number of intermediate care units in which organ-specific problems are treated. Respiratory intermediate care units focus on respiratory failure, in an analogous fashion to coronary care units and stroke units. These wards do not have the complete infrastructure and staff of conventional intensive care units, but are much better equipped and staffed than regular wards. The leadership of individual units depends on the organ in focus, since specialised knowledge is essential for successful patient management.

The development of specialised weaning centres and long-term care facilities for patients who cannot be weaned from ventilation has important economic advantages as every long-term ventilated patient in a regular intensive care unit reduces the capacity for surgical operations, which, inevitably, are an important source of finance for hospitals. Specialised units for long-term ventilation, sometimes outside hospitals, are likely to develop further; they will care not only for the above-mentioned patients with chronic ventilatory failure, but also for patients discharged from intensive care units who cannot be weaned, and who need long-term invasive or noninvasive ventilation.

Important recent developments in intensive care include devices for extracorporeal oxygenation and removal of CO₂. Extracorporeal membrane oxygenation is becoming available in highly specialised centres. CO₂ removal systems are already being applied in weaning and might be useful in respiratory intermediate care units as a short-term ‘bridge’ in patients with acute on chronic respiratory failure, e.g. in patients with AE-COPD. The development of permanent extracorporeal lung replacement for the treatment of chronic respiratory insufficiency appeared to be unrealistic until

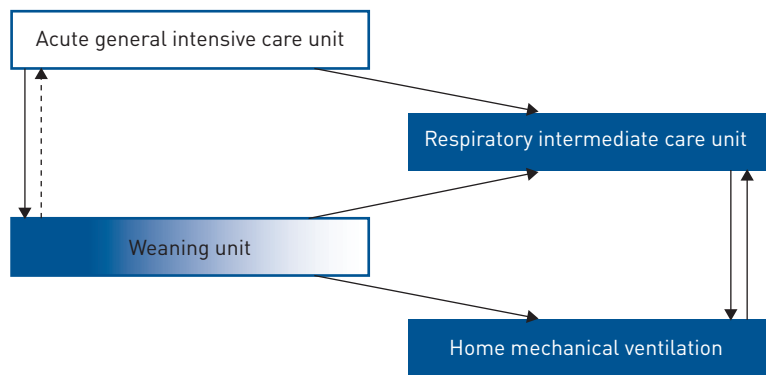


Figure 2 – Levels of care for patients with respiratory failure. Aspects shaded in blue are led by respiratory physicians.

recently, but with improvements in membranes, miniaturised pump systems, and longer-lasting catheters, the artificial lung has become a realistic prospect. Intensive care medicine is thus generating new challenges for respiratory physicians, which extend their range and for which educational programmes will be necessary in order to meet the growing requirements.

As mentioned previously the organisational models of intensive care medicine differ considerably between, and sometimes within, European countries, with variation in personnel, educational standards, infrastructure and responsibilities. The overall relationship between respiratory medicine and intensive care medicine across Europe is unclear, apart from in the field of domiciliary ventilation. In some countries, respiratory medicine is only partly involved in respiratory intermediate care units, weaning centres and the initiation and management of home ventilation. Similarly, respiratory medicine is still under-represented in conventional intensive care and the subject is not part of the respiratory physician's curriculum in all countries. Several training programmes have been developed to harmonise educational and training core curricula in intensive care in Europe. The European Society of Intensive Care Medicine (ESICM) has produced the Patient-centred Acute Care Training curriculum, an up-to-date, online, modular curriculum for intensive (critical) care medicine. This is an educational resource aimed at advancing and harmonising the quality of acute and critical care medicine training and practice. The European Board of Anaesthesiology, under the auspices of the Union Européenne des Médecins Spécialistes, has developed the Anaesthesiology, Pain and Intensive Care Medicine Curriculum. Additionally the ESICM has developed the European Diploma in Intensive Care, while the European Society of Anaesthesiology has developed a European Diploma in Anaesthesiology and Intensive Care.

A respiratory critical care syllabus has been developed and published under the European Respiratory Society HERMES initiative (see chapter 36) and this will be followed by a specific curriculum and diploma. Quality indicators for staff and infrastructure need to be refined for all areas of intensive care medicine (full intensive care units, respiratory intermediate units, weaning centres, home ventilation). Respiratory physicians should be responsible for defining the criteria for the last three.

Respiratory medicine should also contribute to the standards and curricula for other professional groups involved in intensive care medicine and should be the lead speciality for defining educational standards for respiratory intermediate care units, weaning units and home ventilation.

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Introduction



Key points

- Thoracic surgery has changed profoundly in the 21st century. Multidisciplinary treatment is evolving and surgeons have major collaborative roles in management of lung cancer, respiratory infections, chest trauma, paediatric respiratory disorders and end-stage respiratory insufficiency.
- Today, lung cancer is the most frequent indication for thoracic surgery.
- Lung transplantation is increasingly an option for patients with end-stage lung disease.
- The organisation of thoracic surgery varies between European countries and precise figures on the number of centres and the number of procedures carried out each year are not available.
- Thoracic surgeons need to be equally competent in pre-operative investigation and planning, the surgery itself and post-operative management.
- Recent developments in thoracic surgery have focused on reducing invasiveness, particularly through the use of video-assistance, and on parenchyma-saving techniques such as sublobar resection.
- To further stimulate progress in general thoracic surgery and to improve outcomes for patients, cooperation with respiratory physicians is of utmost importance.

Thoracic surgery has existed as a specific surgical discipline for more than a century. Initially, its main focus was surgery for tuberculosis and bronchiectasis. However, since 1940, rapid progress has been made in surgery for lung cancer, of the oesophagus and, most spectacularly, of the heart. After 1960, cardiac surgery became a separate subspecialty with an emphasis on coronary bypass surgery, valve surgery and congenital heart surgery. In most European countries, general thoracic (noncardiac) surgery is now well demarcated and exists as a separate specialty. However, the number of centres performing thoracic surgical interventions in Europe is unknown and a substantial number of thoracic surgical procedures are still performed outside dedicated thoracic surgical units. Consequently, no accurate figures on the total number of operations are currently available. As an example, thoracic surgery is not a specifically defined entity in Belgium, where it falls within the discipline of general surgery, together with abdominal, cardiac, vascular, paediatric and trauma surgery. Approximately 2000 pulmonary resections are performed each year in Belgium but only a minority of centres carry out more than 50 major thoracic operations per year. The other operations are spread widely over smaller centres performing less than 10 interventions yearly.

In some other countries (for instance the UK), thoracic surgery is part of the framework of cardiothoracic surgery. In Germany, large

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Lung transplantation is increasingly an option for patients with end-stage lung disease

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free-standing units exist in combination with respiratory medicine, together with smaller cardiothoracic units and nonspecialised thoracic surgery units within general surgical clinics. Clearly, therefore, there is still no uniformity regarding general thoracic surgery in Europe. To define a more precise structure for general thoracic surgery, a working group has been established by the European Association for Cardio-thoracic Surgery (EACTS) and the European Society of Thoracic Surgeons (ESTS).

Recently, the Union Européenne des Médecins Spécialistes (UEMS) has created a specific thoracic surgical division, related to its general and cardiothoracic surgical sections. The division's statutes were finalised in June 2012 and representatives from each European country will be nominated. Specific criteria for training and accreditation in thoracic surgery are being developed.

The UEMS division of thoracic surgery is currently surveying the practice of thoracic surgery throughout the European Union (EU). At the time of writing, responses have been received from 22 EU member states: in 11, thoracic surgery is a separate single specialty; in eight, it is considered part of cardiothoracic surgery; and in three, there is no separate specialty of thoracic surgery.

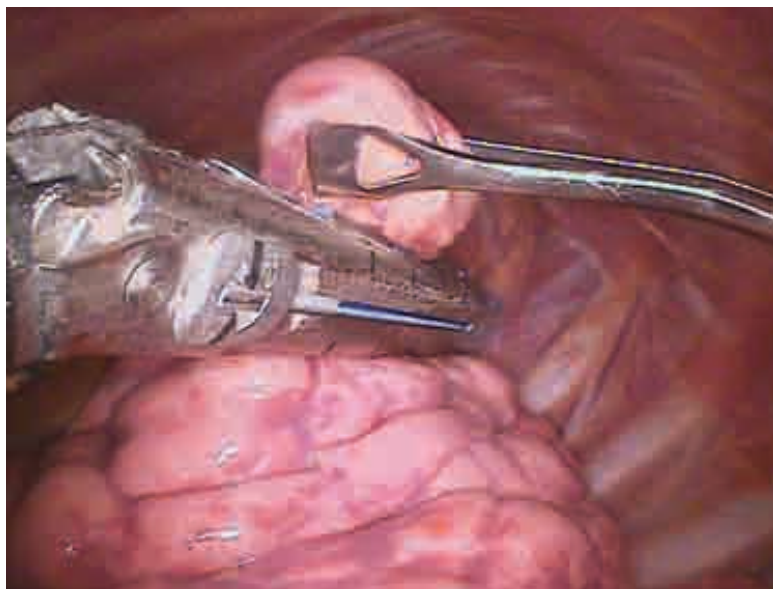
To obtain more precise data on the number of general thoracic surgical procedures carried out in Europe, several large databases have been created. The ESTS has established a voluntary database for general thoracic surgery. In 2011, a total of 24 574 lung resections were reported: lobectomy (removal of a lung lobe) represented 57.5% of cases and pneumonectomy (removal of a lung) 9.5%. A total of 16 710 cases of primary lung cancer were reported, lobectomy and bilobectomy being performed in 76% of cases. Most submitted cases originate from France, where a national database is already established with contributions from 90 thoracic surgical units. The Second National Thoracic Surgery Database Report of the Society for Cardiothoracic Surgery in Great Britain & Ireland, published in 2011, records an impressive total of 109 388 primary lung cancer resections, performed between 1980 and 2010 – with a significant increase in the rate from 2005 onwards.

Standards of care

By definition, general thoracic surgery includes the knowledge, technical skill and judgement required to diagnose and treat diseases of the chest. The entire spectrum

comprises the chest wall, pleura, lungs, trachea and bronchi, mediastinum, diaphragm and oesophagus, in adults and children. General thoracic surgery requires in-depth knowledge of physiology, imaging, organ function testing, investigation, pre-operative evaluation, post-operative and critical care, trauma, oncology and transplantation. It also includes experience in multidisciplinary treatment protocols. The main competence of a thoracic surgeon is the pre-,

Figure 1 – Wedge excision of lung apex by video-assisted thoracic surgery in a patient with pneumothorax.



Standard

- Lobectomy
- Bilobectomy
- Pneumonectomy

Conservative or lung parenchyma-sparing operations

- | | |
|----------|--|
| Proximal | Bronchotomy |
| | Rotating bronchoplasty |
| | Bronchial or tracheal wedge excision |
| | Bronchial or tracheal sleeve resection |
| Distal | Segmentectomy |
| | Wedge excision |

Extended procedures (lung + other structure)

- Pericardium (intrapericardial pneumonectomy)
- Diaphragm
- Chest wall (ribs, vertebrae)
- Superior sulcus (Pancoast tumour)

Table 1 – Types of operative procedures.

intra- and post-operative care of patients with general thoracic surgical diseases. This includes the investigation of patients, decisions on specific treatment, performance of technically correct procedures and provision of expert post-operative care.

Techniques and procedures

Since 1990, less invasive techniques have been developed, with video-assisted surgical procedures replacing the classical thoracotomy for some indications. The percentage of these minimally invasive procedures has gradually increased over recent years: in the UK, approximately one-third of all lung resections are now performed by video-assisted thoracic surgery (VATS). Similar trends are found in other countries. (An example of a subtotal pleurectomy performed by VATS can be seen at the Multimedia Manual of Cardio-Thoracic Surgery, dx.doi.org/10.1093/mmcts/mms008). A VATS wedge excision is illustrated in figure 1. Recently, robotic surgery has also been introduced in thoracic surgery, providing a superb three-dimensional view and allowing precise operative interventions using highly flexible robotic arms. Thymectomy and resection of smaller anterior mediastinal tumours can be easily accomplished by robotic surgery. Lobectomy and, to a lesser extent, pneumonectomy remain the classic surgical procedures for lung cancer (table 1). However, over the past two decades major efforts have been made to develop more lung parenchyma-saving interventions. Sleeve and double-sleeve lobectomies using broncho-/tracheoplastic and vascular reconstructive techniques have been introduced successfully. These techniques avoid the need for pneumonectomy, resulting in lesser impact on pulmonary function and thus better quality of life, or allowing for appropriate resectional surgery in patients who otherwise wouldn't tolerate pneumonectomy. Increasing attention is also directed towards sublobar resection, *i.e.* anatomical segmentectomy or wide wedge excision in small peripheral cancers: results so far are promising, with similar survival figures in nonrandomised studies to those obtained after classic lobectomy. Indications for surgical treatment of nonsmall cell lung cancer are listed in table 2.

Lung metastasectomy has become a well-accepted procedure in the treatment of lung metastases originating from some solid-organ tumours.

Although still controversial, radical surgery in malignant pleural mesothelioma consisting of either extrapleural pneumonectomy or extended pleurectomy/decortication with or without resection of the diaphragm and pericardium, is performed by a number of thoracic surgeons in an effort to obtain maximal tumour reduction as part of a multimodal therapeutic strategy.

Lung transplantation

New developments and multidisciplinary cooperation have increasingly made lung transplantation a valid option for selected patients with end-stage lung disease. Worldwide transplantation activity has increased year on year over the past 25 years

Within the seven-country Eurotransplant community, 1182 lungs (528 double lung, 89 single lungs and 37 lungs plus other organ) were transplanted in 2011, an increase of 6.6% compared to 2010. The most common indications are end-stage emphysema, cystic fibrosis, idiopathic pulmonary fibrosis and pulmonary hypertension. Double lung transplantation has become the standard. According to the most recent adult lung transplant report of the International Society for Heart & Lung Transplantation (ISHLT), 3519 lung transplantation procedures were performed globally in 2010, the highest



Definite

Stage IA	T1a,bN0
Stage IB	T2aN0
Stage IIA	T2bN0, T1a,bN1, T2aN1
Stage IIB	T2bN1, T3N0
Stage IIIA	T3N1

Investigational

Stage IIIA	T1–3N2, T4N0,1
Stage IIIB	T4N2, T1–4N3

Exceptional

Stage IV – single metastasis
Stage IV – multiple metastases

Table 2 – Indications for surgical treatment of nonsmall cell lung cancer. See chapter 19 for an explanation of lung cancer staging.

number ever reported. Although lung transplantation remains a high-risk procedure, survival results have improved over the past decade. Five-year survival is now about 50% overall, but more experienced centres now regularly report 5-year survival figures in the region of 70% (figure 3). However, long-term survival remains low compared with transplantation of other solid organs, due to chronic infection and rejection leading to bronchiolitis obliterans syndrome (BOS), which remains the Achilles’ heel of lung transplantation.

Organisation of thoracic surgical centres

To ensure the best possible patient care in thoracic surgery, the EACTS/ESTS working group states that it should be performed within the logistical and economic framework of specialised units. These units should be designed to allow patient care and treatment according to recommended

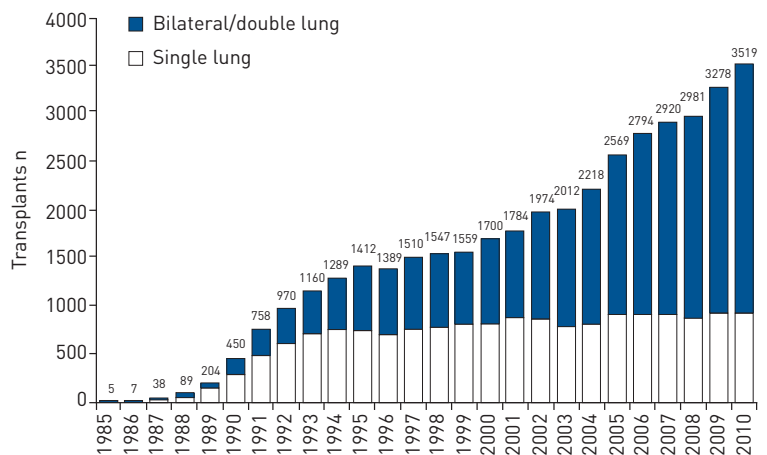


Figure 2 – The global number of adult lung transplants reported to the International Society for Heart & Lung Transplantation each year since 1985. Reproduced from CHRISTIE *et al.*, 2012, with permission from the publisher.

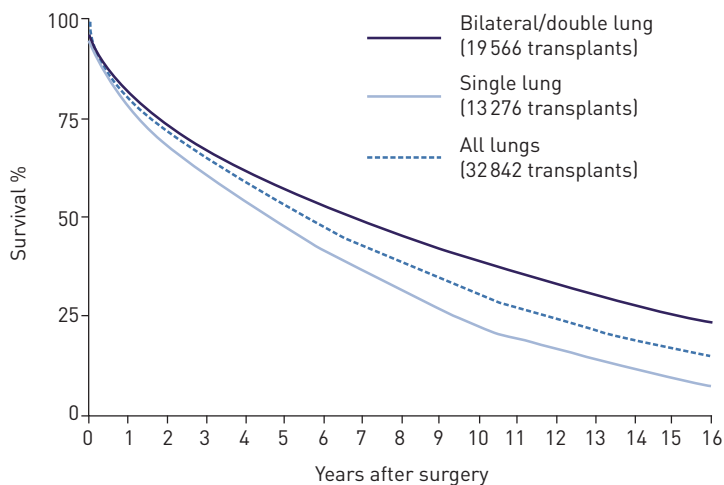


Figure 3 – Survival of adult lung transplant patients (operations performed between 1994–2010) according to type of transplantation. Reproduced from CHRISTIE *et al.*, 2012, with permission from the publisher.

standards, as well as education of surgical trainees, continuous development and research in thoracic surgery.

The working group proposed two types of thoracic surgical centres: highly specialised centres within, or associated with a university, performing at least 250 major thoracic procedures per year, and standard units which are free-standing or combined with cardiac, vascular or general surgery. In a standard unit at least 100 major interventions should be performed annually. Lung transplantation and its alternative procedures should be performed only in centres with special interest and with cardiac surgical facilities.


Changing roles

Thoracic surgery has changed profoundly in the 21st century. Multidisciplinary treatment is evolving and thoracic surgeons have become major team players in diseases related to thoracic oncology, infection, trauma, paediatric disorders and end-stage respiratory insufficiency.

The changing pattern of thoracic surgical practice is exemplified by a recent new classification of adenocarcinoma, which will have a profound impact on surgical decision making and treatment. For smaller tumours the role of sublobar resection is reconsidered as mentioned above. In the most recent revision of the Tumour, Nodes, Metastases (TNM) classification for lung cancer, important changes were made in the T and M descriptors, with definition of new subcategories. A new lymph node map was introduced together with the concept of nodal zones, reconciling previously published lymph node classifications. A prospective database has already been created to refine the classification ahead of the next revision.

The future

In order to increase the profile of thoracic surgery within the European Union and elsewhere in Europe, further harmonisation in practice and organisation is necessary. This relates not only to training in thoracic surgery, but also to certification of dedicated thoracic surgical units.



Unified databases, to which the majority of thoracic surgeons contribute, should be made available, detailing not only mortality but also specific outcome measures related to morbidity, survival and quality of life. Postgraduate education remains essential to ensure high-quality surgical interventions, as has recently been demonstrated by a study from the Netherlands evaluating completeness of lymph node dissection in dedicated thoracic surgical centres. Thoracic surgeons should be further involved in randomised clinical trials comparing newly introduced treatment modalities, such as stereotactic radiotherapy or radiofrequency ablation, to classical surgical procedures.

As their field is constantly changing, thoracic surgeons should be prepared to adapt to a new environment bringing not only new challenges but also opportunities to further develop and refine this fascinating specialty.

To further stimulate progress in general thoracic surgery, cooperation with respiratory physicians is of utmost importance to improve the outcome for patients. The structure of the European Respiratory Society provides a solid basis for mutual interactions and exchange of knowledge between the different groups of respiratory medicine and general thoracic surgery. Better patient care will be the ultimate result.

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