Definition and scope:

Respiratory disorders in children are common, diverse and largely managed in Primary and Secondary care (the wide range of problems is listed in the National Definitions Set¹). However, tertiary respiratory services are required for the purposes of supporting the local clinicians for complex problems of common disease e.g. difficult asthma, as well as providing full or shared care for rare disease. In addition tertiary services have important links to many other specialties (in particular intensive care, neonatology thoracic surgery, ENT and neurology). Other essential disciplines include specialist nurses, dietetics, physiotherapy, physiological measurement technicians and psychology.

Scotland has realigned services over recent years to focus specialist volume services (see Table A) in larger centres, to transport sick children to appropriate paediatric intensive care facilities and to increase the standard of healthcare for children dependent on technology. Tertiary Respiratory Medicine has expanded to enable these service redesigns and developments, but without any planning or co-ordination. There are large discrepancies in the services that can be provided at each of the tertiary centres.

Table A: Specialist respiratory services provided for large populations

- Bronchoscopy
- Home oxygen and monitoring
- Diagnostic pulmonary function testing
- Management of respiratory complications in neuromuscular disease
- Management of recurrent aspiration problems
- Investigation and treatment of recurrent lower respiratory tract infections
- Diagnosis and treatment of sleep disordered breathing
- Home ventilation

Similar to other services the development of Managed Clinical Networks is seen as the best way of delivering Tertiary Respiratory Medicine across Scotland, while ensuring equity of access. Without substantial investment in resources in Tertiary Paediatric Respiratory Medicine it will not be possible to meet these aspirations or even maintain the current level of service with the impending changes to working patterns set by the European Working Time Directives.

Incidence and prevalence:

See Appendix 1 for detail

Respiratory conditions are the commonest cause of paediatric hospital admission, accounting for 14% of UK hospital admissions² and over 50% of long-term illness in children³. Asthma is the commonest cause of school absence, Cystic Fibrosis the commonest life limiting inherited condition. Table B shows the incidence prevalence of respiratory conditions for a region e.g. Grampian or Tayside, with 100,000 children aged 0-14 years and approximately 6,000 births per year.
### Table B. Incidence/prevalence of Respiratory conditions

<table>
<thead>
<tr>
<th><strong>Primary chronic conditions</strong></th>
<th><strong>Children affected</strong></th>
<th><strong>Comments</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Asthma</strong></td>
<td>20,000</td>
<td>2% with complex/severe disease</td>
</tr>
<tr>
<td><strong>Cystic fibrosis</strong></td>
<td>40</td>
<td>All require specialist care</td>
</tr>
<tr>
<td><strong>Chronic lung disease of newborn</strong></td>
<td>6 p.a.</td>
<td>Home oxygen required</td>
</tr>
<tr>
<td><strong>Non-CF bronchiectasis</strong></td>
<td>20-30</td>
<td>6 of whom have Primary Ciliary Dyskinesia</td>
</tr>
<tr>
<td><strong>Sleep disordered breathing</strong></td>
<td>100</td>
<td>Detailed assessment required</td>
</tr>
<tr>
<td><strong>Long-term ventilation</strong></td>
<td>2-4</td>
<td>Complex multidisciplinary care package needed</td>
</tr>
<tr>
<td><strong>Rare serious lung disease</strong></td>
<td>2-4</td>
<td>Requires specialist diagnostic services</td>
</tr>
<tr>
<td><strong>Congenital lung and airway disease</strong></td>
<td>6 p.a.</td>
<td>Requires specialist diagnostic services</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th><strong>Complications of other conditions</strong></th>
<th><strong>Children affected</strong></th>
<th><strong>Comments</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Neuromuscular</strong></td>
<td>25</td>
<td>Severe disease</td>
</tr>
<tr>
<td><strong>Neurological</strong></td>
<td>100</td>
<td>Cerebral palsy or degenerative conditions</td>
</tr>
<tr>
<td><strong>Immune deficiency</strong></td>
<td>4</td>
<td>Immunology input vital</td>
</tr>
<tr>
<td><strong>Oncology</strong></td>
<td>Unknown</td>
<td>Mostly “emergency” input for respiratory infection</td>
</tr>
<tr>
<td><strong>Chest wall and scoliosis</strong></td>
<td>Unknown</td>
<td>Liaison to plan and monitor around surgery</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th><strong>Acute admissions</strong></th>
<th><strong>Children affected</strong></th>
<th><strong>Comments</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>LRTI</strong></td>
<td>300 p.a.</td>
<td>Often managed by non-specialist</td>
</tr>
<tr>
<td><strong>Croup</strong></td>
<td>100 p.a.</td>
<td>Often managed by non-specialist</td>
</tr>
<tr>
<td><strong>Empyema</strong></td>
<td>4 p.a.</td>
<td>Involves thoracic surgery</td>
</tr>
</tbody>
</table>

Figures base on Scottish single centre audits or published prevalence data

Many common respiratory illnesses can be very well managed in primary and secondary care. However, those children with severe common illnesses e.g. asthma or pneumonia, or with rare respiratory disease require the involvement of a specialist multidisciplinary team.

The prevalence of asthma has increased over the last 20 years but there is evidence that this may have peaked. However, the workload of seriously affected children, especially in adolescence, may have continued to increase. In these individuals the delicate balance of benefits to risks of steroid therapy requires specialist input. Cystic fibrosis prevalence is likely to increase with the introduction of new-born screening in 2003. The incidence of TB is already increasing with changes in ethnic minority groups in Scotland. Changing expectations from parents and professionals are increasing the demand for more intensive respiratory intervention e.g. non-invasive ventilation for Duchenne Muscular Dystrophy. It is now recognised that several conditions, such as Primary Ciliary Dyskinesia, non-CF bronchiectasis and sleep disordered breathing (especially Obstructive Sleep Apnea in obese children) are currently underdiagnosed, leading to preventable morbidity. A further influence on workload is the presence of new cohorts of children who have survived previously fatal problems but with respiratory sequelae e.g. extreme prematurity.
Mapping of current services:

See appendix 2 for details

Tertiary respiratory services are currently provided in Aberdeen, Dundee, Edinburgh and Glasgow with varying levels or staff. The presence of PICU (Edinburgh and Glasgow) is a significant influence on the service, particularly with respect to national services and ventilatory support. For some diseases, such as CF, these inequalities are being increasingly recognised. However, for other patient groups, who do not have strong support groups, their equity of access to high quality respiratory services remains unmet. Clinicians find it difficult to commission and fund these low volume, but complex and high cost services, which are often established on an ad hoc basis, without planning or co-ordination.

As outlined above, Tertiary Respiratory services are dependent on many other specialties and high quality care requires a multidisciplinary team approach. The current national shortage of trained paediatric radiologists is of great concern as high quality imaging is an essential aspect of assessment and diagnosis.

The possibility for shared care arrangements in Scotland is good, with informal links already established e.g. Scottish Paediatric Respiratory Interest Group (SPRING). General paediatricians, with a significant respiratory interest, provide excellent levels of care in secondary centres. However, this provision is patchy and consideration should be given as to a method of providing a more even distribution of good quality respiratory care throughout Scotland (in terms of both medical and AHP care).

No single centre in Scotland will be able to provide a comprehensive out-of-hours service within European Working Time Directives.

Review of current practice:

See Appendix 1 for details

Current practice is that the four centres work independently, with close links for Cystic Fibrosis practice than for other disease groups. SPRING has enabled a framework for collaboration and discussion. The benefits of this group are already being seen in terms of service planning, research development and document sharing. It is agreed that many tertiary respiratory services should be provided in a number of centres in Scotland, and that some specialist services should be provided at only one or two centres e.g. Home ventilation. Tertiary Respiratory Medicine does not lend itself to a single centre solution, because of the higher population prevalences of respiratory illness relative to other, more tightly focused, specialist services.

The development of the new Royal Aberdeen Children's Hospital, the Tayside Institute of Child Health and considered developments in Edinburgh and Glasgow provide an excellent opportunity for service redesign, with modern, coherent provision of tertiary services within a dedicated, designed for purpose space. In addition the existing strong links between University Child Health Departments and the clinical service will foster pioneering clinical research both locally and for collaborative multicentre projects.

There are 3 areas of particular concern:

1. Sleep disordered breathing and home ventilation services. The demand is increasing rapidly and the increasing prevalence of obesity is leading to increased numbers of children with...
obstructive sleep apnea. Current services are patchy at best and lack the appropriate infrastructure support. It is likely that national guidelines to be published soon will highlight a considerable service gap.

2. Respiratory care of children with neurological disorders. Changing expectations and demonstration of gains in quality and quantity of life has led to an increase in the intensity of respiratory support provided. Children with chronic severe disability are living longer, occupying more intensive care beds, and are becoming more technology-dependent. Respiratory paediatricians are asked to lead the provision of care for these patients, but without adequate resources to do so.

3. Diagnostic services. Appropriate, standardised, equipment with fully trained technical staff is crucial to any tertiary service. Investment in IT would allow investigations to take place locally, thereby reducing unnecessary travel for patients. Flexible bronchoscopy is established in 3 centres but large amounts of time/effort are wasted as no centre has dedicated theatre sessions or supporting staff.

The transition into the respiratory service for those with neonatal conditions such as chronic lung disease is inconsistent, with some home oxygen services provided by neonatologists and some by respiratory paediatricians. Hospital readmission is common and it is imperative that a seamless service is provided for these patients. The transition between paediatric and adult respiratory services is also very variable. CF generally has good transitional arrangements, probably because of the existence of dedicated relatively well funded adult services. In other conditions e.g. asthma, immune deficiency, congenital lung disease, etc. good transition arrangements are not established.

It is disappointing that recent NHS Scotland pronouncements on Oxygen therapy appear to have overlooked paediatric aspects of care. Children may need this service in fewer numbers but the health gain is relatively large.

**Workforce planning and Training:**

**Workforce**

The current distribution of Tertiary Respiratory Workforce is provided in Appendix 2.

There are 5 WTE clinical tertiary respiratory paediatricians working in Scotland caring for a population of c 5 million (800,000 children). Four of the 11 individuals contributing to respiratory paediatrics are university staff. To provide an adequate level of service, on the basis of current workload patterns, it is estimated that an additional 6 WTE Tertiary Respiratory Paediatricians will be required. This is formulated from the limited input available from University members of staff and information from current workpractice diaries.

The additional workforce required to support a tertiary respiratory service is provided in Appendix 3. This calculation is based on recommended standards, and the need to establish a Scotland-wide clinical network.

**Training**

- Medical. Accredited tertiary respiratory training is available in Edinburgh and Glasgow with partial training in Dundee and Aberdeen. Allowing for retirement and some loss of trainees to other regions, it is estimated that there needs to be 2 SpRs training per year, for the next 5 years, to maintain the current service. If the service is to expand to a number of WTE consultants sufficient to provide a comprehensive country-wide service then this level of training needs to continue for 10 years before there is a requirement to curtail training posts.
AHPS and other health care professionals. Edinburgh RHSC runs a Degree Nursing Module for Paediatric Respiratory Medicine. However, many specialist asthma nurses can only gain their training by distance learning, often funded by pharmaceutical companies. Training courses in sleep medicine are available in USA but the only ones in the EU cover polysomnography only. The courses in the USA are expensive. There are no known courses in paediatric respiratory physiotherapy, dietetics, or physiological testing, although many of the manufacturers of lung function testing equipment provide local training.

Quality standards/outcome indicators:

Standards of care for asthma are defined in the SIGN/BTS guidelines, and NICE guidance on inhalers. The National Asthma Campaign has recently published a Patient's Charter. Research work is in progress between Dundee and Glasgow that may help inform the debate on standards of care in asthma, but there is a need for a national database. Currently all Scottish centres participate in the UK asthma audit annually. The CF Trust has published on standards of care. In addition the UK CF database (based in Dundee) enables outcomes to be assessed. There are guidelines for long-term ventilation and The Scottish Ventilation Support database is at an advanced stage of development, which will enable annual reviews, and to aid the care. European standards exist for Primary Ciliary Dyskinesia, and for respiratory care in neuromuscular disorders. International standards have been published for the management of sleep-disordered breathing, and UK standards are being developed.

Review of Education and Information:

On a national scale, there are 2 annual Cystic Fibrosis and 1 Asthma education days for those in primary, secondary and tertiary care. Medical staff are research active and participate in national (UK) and international meetings presenting local projects and as invited speakers. Similarly, nursing and AHP staff are involved in education and training, not just in Scotland. The recently established Scottish Paediatric Respiratory Interest Group (SPRING) has developed a program of quarterly educational meetings open to Scottish medical and nursing staff with a respiratory interest. Glasgow has developed an asthma booklet for parents and families that will probably be disseminated for wider use. The framework exists for sharing such educational initiatives in other disease areas. There is a tendency to leave lay education to the lay groups such as the National Asthma Campaign and Cystic Fibrosis Trust who have excellent information literature.

Identify current research:

See Appendix 4 for details.

With one of the highest prevalence of paediatric asthma in the world, a geographically defined population and excellent research-based Universities, Scotland is well placed for first-rate paediatric respiratory research. However, there are limited opportunities for clinical research. Below is a summary of current research interests:

- **Aberdeen**: Professor Peter Helms, Professor of Paediatrics (Asthma genetics and epidemiology); Dr Stephen Turner, Senior Lecturer (Infant lung function testing, exhaled nitric oxide). Dr Richard Brooker (inflammatory response to RSV infection)
- **Dundee**: Professor Richard Olver, Professor of Child Health (Lung development, epithelial ion and fluid transport), Dr Somnath Mukhopadhyay, Senior Lecturer (nutrition and lung growth, adherence studies in asthma and CF, genetic studies in asthma), Dr
Anil Mehta, Senior Lecturer (CFTR regulation, UK CF Database, CF genetics and epidemiology).

- **Edinburgh**: Dr Steve Cunningham, Part Time Senior Lecturer (Oxygen toxicity on newborn lung, Non-invasive markers of airway inflammation). Dr Tom Marshall (CF clinical trials)
- **Glasgow**: Dr James Paton, Reader (Compliance in asthma therapy, Exercise physiology, care pathways in asthma), Dr Neil Gibson (Daytime somnolence in OSAHS). Dr Anne Devenny (CF clinical trials), Dr Dominic Cochran (Asthma clinical trials)

**Implications for Stakeholders:**

- **Patient Stakeholders**: Respiratory services in Scotland, lack coherence and common pathways: Asthma information, protocols and access to tertiary opinion are limited; CF patients have significant inequality of access and paediatric CF has lacked central support; Sleep services are scanty given the evidence for long term patient benefit; Access to non-invasive respiratory support is too restricted (for a low cost option that increases life expectancy by 33% in some disease groups).
- **Primary Care Stakeholders**: Tertiary Respiratory Services can provide primary care with support for the most common primary care consultations and, with protocol development help, guide review of those patients who may need an alternative assessment.
- **Secondary care Stakeholders**: Those in secondary care need more support, assistance with guidelines and rapid, good quality assessment and opinion. The rapid change of information and patient expectation requires tertiary support, if Scotland’s many (and excellent) secondary care centres are to continue to provide for patients locally. If patients are to be cared locally on a 24/7 basis, then a national on-call service should be considered.
- **Tertiary respiratory services**: deliver secondary care for their locality as well as tertiary care for local population and referring units. These centres are under increasing pressure from providing an increasing quality of service, to a wider population, without an increase in funding. Development of out of hours on-call, would require significant investment to enable rationalisation of University staff to do research, and a core team of complex respiratory specialists to be available out of hours for opinion.
- **The existence of SPRING and the Scottish Cystic Fibrosis Group provides for shared planning and discussion of protocols and care pathways.**

**Conclusions and Recommendations for Change**

The current tertiary respiratory services in Scotland have developed ad hoc according to local enthusiasm and needs. There is, therefore, an inequality of service provided to patients. It is clear that the service must change to meet the challenges of the 21st century and to ensure equity of access for children throughout the region.

The recommendations to secure and develop the tertiary respiratory services fall into 3 main areas:

1. **Managed Clinical Networks.** It has been clear for some time that this is by far the best way to deliver tertiary services to a geographically spread-out population. However, they must be adequately resourced or they will fail to deliver. A proposal for a CF MCN was submitted in 2002 but to date there has been no progress in realising this development. Complex respiratory disease would be amenable to a similar network. However, other services such as sleep disordered breathing would be more appropriately centred on
Edinburgh and Glasgow with partial sleep services elsewhere. Sleep disordered breathing requires ventilatory support services and the co-location with PICU. Home ventilation should also be centred on Edinburgh and Glasgow. Rare disease, such as Primary Ciliary Dyskinesia, should be concentrated on 1 centre. Glasgow already provides a diagnostic service whose equipment needs updating to allow collaboration with the leading UK centres.

2. **Diagnostic services.** Flexible bronchoscopy is available in 3 of the centres and requires some realignment (Glasgow takes referrals from Tayside and is busy whereas Aberdeen is under-capacity). Considerable amounts of time/effort are currently wasted because of the lack of dedicated theatre time and support. Lung function testing should be available in all centres with appropriately trained staff and facilities. Full polysomnography is appropriate for Edinburgh and Glasgow. Investment in IT and harmonising of equipment throughout Scotland would reduce unnecessary patient traveling.

3. **Out-of-hours service.** It is clear that even the largest centre (Glasgow) will not be able to provide a 24/7 tertiary respiratory service. Fortunately there is a small demand for out-of-hours direct tertiary respiratory input e.g. flexible bronchoscopy, and consideration should therefore be given to developing an on-call rota so that expert advice is available at all times throughout Scotland. This would be greatly facilitated by improved IT support.

It is clear to all those involved in tertiary respiratory medicine that the current service will not be sustainable in the future without some increase in resources. However, with such an investment the SEHD could make a significant impact and improve the healthcare for children with respiratory illnesses, the commonest cause for UK hospital admissions and long-term illness children. Real improvements for these children and their families can be achieved.

**References:**
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12. Thoracic Society of Australia and New Zealand and Australasian Sleep Association. Accreditation of sleep disorder services, including standards of paediatric laboratories. 2002