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GUIDELINE PAPER

International Primary Care Respiratory Group (IPCRG) Guidelines: Integrating diagnostic guidelines for managing chronic respiratory diseases in primary care

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Summary This is the first of the IPCRG Guideline papers. The IPCRG took on the task of producing an integrated guideline for the management of chronic respiratory diseases in primary care. This included some original work aimed at developing questionnaires suitable for improving the diagnosis and recognition of respiratory diseases in the primary care setting. This paper provides the background evidence and rationale for the integrated diagnostic section of the IPCRG Guideline. It focusses on identifying the unique challenges in respiratory disease diagnosis in primary care, the use of epidemiology to build a co-ordinated diagnostic framework, and the development of tools to support guideline implementation.

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Introduction

The International Primary Care Respiratory Group (IPCRG) took on the task of producing an integrated guideline for chronic respiratory diseases, to be drawn from existing guidelines produced by Allergic Rhinitis and its Impact on Asthma (ARIA) [1], the Global Initiative for Asthma (GINA) [2], and the Global Initiative for Chronic Obstructive Lung Disease (GOLD) [3]. Since each of these existing

guidelines addresses a single disease, one key challenge was to coordinate diagnosis across all three disease states. Diagnosing chronic respiratory disease in the primary care setting can be difficult in itself, and the task of integrating existing guidelines for three disease processes poses special challenges.

Another important task was to adapt guidelines for implementation in the primary care setting. Implementing guidelines can be difficult, and general practitioners (GPs) can often benefit from tools and simplified algorithms to enable them to adapt and implement guidelines while meeting the demands and limitations of busy primary care practice settings.

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This report provides the evidence and rationale for the integrated diagnostic section of the new guideline [4]. We will focus on: (1) identifying unique challenges in respiratory diagnosis; (2) using epidemiology to build a coordinated diagnostic framework, and; (3) developing tools to support guideline implementation.

Unique Challenges of Respiratory Diagnosis

Overlap between allergic rhinitis and asthma

There are important epidemiological and pathophysiological links between allergic rhinitis and asthma, suggesting a strong relationship between the two [1]. The current view is that both upper and lower airways are affected by a common inflammatory process. For this reason, the two conditions should be considered together. Other upper respiratory conditions may occur in conjunction with allergic rhinitis, thus complicating diagnosis. These include sinusitis, conjunctivitis, nasal polyposis, and otitis media.

Underdiagnosis of asthma

Underdiagnosis of asthma may be attributed to a variety of factors, including lack of patient awareness of the disease, reluctance to seek health care in response to respiratory symptoms, and underdiagnosis by GPs [5]. In a general practice setting, asthma diagnosis can be difficult because patient symptoms are intermittent and often nonspecific and because there is a spectrum of clinical presentations [2]. By definition, patients with asthma often demonstrate wide fluctuations in symptoms, and variations in lung function and response to medication. As a result, patient presentation may vary between overt signs and symptoms of the disease or none at all. The boundaries for distinguishing asthmatic patients from those without asthma are not absolute [6], and there is no single test or pathognomic feature which is definitive for the presence or absence of asthma [7]. Asthma is typically diagnosed in the clinical setting based on a combination of history, physical examination, and physiologic testing (i.e., lung function), often over a period of time. Patients with asthma are often misdiagnosed as suffering from recurrent upper or lower respiratory tract infections; therefore, a thorough perusal of the past medical record, together with detailed

history-taking, may often provide additional evidence to support a diagnosis of asthma [8–10].

Asthma in young children

The diagnosis of asthma in infants is difficult. The largest proportion of children under 5 years of age with wheezing are transient wheezers who do not have asthma. In many cases, it is not possible to discriminate between transient wheezers and asthma at an early age, although a positive family history of atopic disease, presence of eczema in the child, and a good response to bronchodilators and/or corticosteroids are more suggestive for asthma. The differential diagnosis of asthma in young children is considerable and includes bronchiolitis, recurrent ENT infections, gastro-oesophageal reflux, cystic fibrosis, congenital anomalies of the airways/lungs, and congenital heart defects. Young children whose primary symptom is recurrent or persistent cough, or who wheeze with respiratory infections, are often misdiagnosed as having bronchitis or pneumonia and thus ineffectively treated with antibiotics or cough suppressants [2]. Diagnosis is further complicated by the difficulty of performing lung function testing in young children.

Occupational asthma

Workers exposed to inhalant chemicals or allergens in the workplace can develop asthma and may be misdiagnosed as having chronic bronchitis or chronic obstructive pulmonary disease (COPD). Early recognition, strict avoidance of further exposure, and early treatment are essential [2]. Diagnosis of these patients may require help from a specialist [11].

Underdiagnosis of COPD

Underdiagnosis of COPD is common as this disease is often not recognised and diagnosed until it is moderately advanced [3]. Since COPD develops slowly, patients with early disease may have few symptoms; therefore, they may not seek medical help, and their doctor may not recognize that they have damaged lungs. Many patients adapt to their symptoms, gradually decreasing their activity level over time, without seeking medical help [12]. Some symptoms associated with COPD, such as cough and sputum production, are nonspecific. It is difficult to measure how often this disease is missed, but the figure of 50% undiagnosed cases is commonly cited [13]. Many of these undiagnosed cases are at an

early stage of disease when efforts to halt disease progression would be most effective.

Underuse of spirometry in primary care

One of the barriers to the diagnosis of COPD is the difficulty with which spirometry is performed in the general practice setting. Limitations in equipment and time contribute to the fact that primary care physicians rarely use spirometry to detect COPD, as recommended in the guidelines [14–19].

Overlap between COPD and asthma

Overlap between the clinical pictures of asthma and COPD commonly leads to problems in distinguishing these two conditions. Patients with COPD are often incorrectly labelled as having asthma [20,21]. Accurately distinguishing between asthma and COPD is recognised as a major diagnostic challenge in the clinical setting. Shortness of breath is a common feature of both conditions, and it can be difficult to distinguish asthma and COPD in some patients [22–25]. In the case of patients with combined asthma and COPD, the degree of reversibility in airflow obstruction alone may be insufficient to establish a clear diagnosis. In older adults, this problem is particularly acute. The National Asthma Education and Prevention Program in the USA issued a report highlighting special considerations for managing asthma in the elderly [26]. The report noted that some elderly patients display characteristics of both asthma (i.e., some reversibility of airway obstruction) and COPD (i.e., some degree of nonreversible obstruction), thus creating diagnostic confusion. The report also addressed the need to differentiate asthma from other conditions seen with increased frequency in older populations, such as interstitial lung disease, bronchiectasis, cardiac disease (such as angina and congestive heart failure), upper airflow obstruction, pulmonary embolism, bronchogenic carcinoma, aspiration, and gastroesophageal reflux.

Differing guideline approaches

In addition to disease-specific challenges, the existing ARIA [1], GINA [2], and GOLD [3] guidelines employ different approaches to diagnosis. The ARIA guideline indicates that the diagnosis of allergic rhinitis should be based on the history of allergic symptoms and diagnostic testing directed towards the detection of IgE or IgE-mediated allergic reactions [1]. The GINA guidelines emphasize the role of the medical history and physical examination in the diagnosis of asthma,

emphasizing the classic presentation of episodic dyspnoea or chest tightness and findings of expiratory wheezes [2]. Pulmonary function and additional studies are used primarily to confirm the diagnosis. GOLD, on the other hand, begins with symptoms (cough and sputum) and risk exposure (primarily cigarette smoking), but presents spirometry as the primary approach for identifying airway obstruction, providing standardized protocols for interpreting lung function tests [3]. The differences between these approaches exacerbates the diagnostic problems faced by the primary care health professional, who must evaluate patients with consideration of all relevant diseases.

Developing An Epidemiologic Framework

The epidemiology of chronic respiratory diseases lends support for an approach to diagnosis that reflects different clinical presentations based on age demographics.

Allergic rhinitis is unlikely in children below 3–4 years of age [27], and allergen-specific serum IgE tests and allergy skin testing are less reliable in children under the age of three [28]. The prevalence of rhinitis in older children varies widely, from 0.8% to 14.9% in the 6–7 year-old age group and from 1.4% to 39.7% in the 13–14 year-old group, and allergic rhinitis is significantly correlated with asthma in this group [2]. This relationship between allergic rhinitis and asthma continues into adulthood.

Among children, asthma prevalence has been measured at between 0–30% in various populations [2]. Data on asthma prevalence in adults are more controversial, and large variations in asthma prevalence have also been found for adults across different populations. Asthma epidemiology clearly differs for population sub-groups of children, adults, and the elderly, due to changes in lung function, exposure to risk factors, and other factors over the life span. Differential diagnosis considerations also vary widely for these sub-groups, complicating the diagnostic approach. For example, in preschool children, the younger the child the greater the likelihood that something other than asthma explains recurrent wheezing [2]. The age threshold at which a child can actively participate in lung function testing serves as an important demarcator for diagnostic purposes. GINA guidelines recognize the elderly, paediatric, and occupational populations as posing specific challenges with respect to the diagnosis of asthma [2].

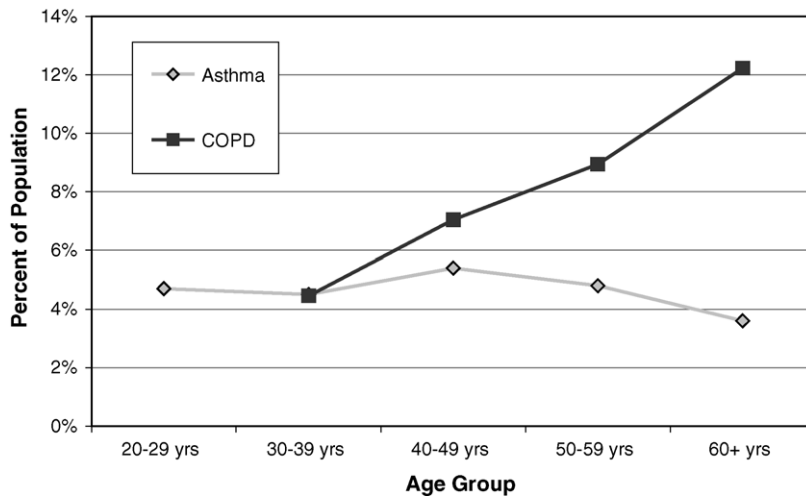


Figure 1 Prevalence of Patient-Reported Asthma and COPD [reproduced from 46].

Nationally representative USA surveys illustrate age-specific prevalence trends in asthma relative to COPD in adults. Data from the National Health Interview Survey show that the prevalence of self-reported asthma peaks in the 12–17 year old age group at 14.2% [29], drops to 9.2% among 18–44 year olds, and continues to decrease gradually with age [30]. At the same time, the prevalence of emphysema and chronic bronchitis begins a steady increase, starting from rates of 0.3% and 1.5% respectively in 18–44 year olds. Data from the Third National Health and Nutrition Examination

Survey illustrate the rise in patient-reported COPD with age (Figure 1) [31,32]. Among adults aged 30–39 years, COPD is no more common than asthma; COPD prevalence then rises steadily with age until, over the age of 60, COPD is three times more common than asthma. When airway obstruction is considered independent of diagnostic label, the rise with age is even more pronounced [32].

Taken together, this evidence suggests discrete diagnostic groups, each of which require a tailored diagnostic approach (Table 1).

Table 1 Chronic Respiratory Diseases: Principal Epidemiologic and Diagnostic Issues

Age Group	Primary Epidemiologic Issues	Primary Diagnostic Issues
Young children (under age 5–6)	Asthma not the most common cause of chronic respiratory symptoms Allergic rhinitis is uncommon under age 4	Exclude competing causes of chronic respiratory symptoms Lung function testing not available
Older children (age 6 and older)	Asthma and allergic rhinitis are the most common causes of chronic respiratory symptoms	Document reversible obstruction Consider coexisting atopic disease
Younger adults (up to age 40–45)	Asthma and allergic rhinitis are the most common causes of chronic respiratory symptoms Occupational exposures may be important	Document reversible obstruction Consider coexisting atopic disease Identify occupational contribution (if any)
Older adults (age 40 and older)	COPD is more common than asthma, but may be asymptomatic Characteristics of COPD and asthma may overlap	COPD case-finding Differentiate asthma from COPD Identify occupational contribution (if any)

Developing Tools to Facilitate Diagnosis in Primary Care

Guidelines are difficult to implement, and dissemination alone is insufficient to guarantee uptake [33,34]. Lowering barriers to acceptance can help. Approaches include summary algorithms that guide practitioners through the key points of a guideline, and support tools to help simplify key decisions. Although these types of tools are more often designed to aid treatment decision-making, the same approach may be applied to diagnosis.

Diagnostic algorithm

For the IPCRG guidelines, a diagnostic algorithm was based on the age groups and primary diagnostic issues described above. As with all guidelines, this algorithm represents an oversimplification of the complex diagnostic issues at hand. Nevertheless, it provides an integrated approach to diagnosis, and creates a logical flow of patients through a busy practice.

Case-finding Tools

Since respiratory symptoms are among the most common presenting complaints in primary care, it is impractical to investigate fully every patient with symptoms. It is more helpful to focus on patients most likely to benefit from a full diagnostic evaluation. Screening and case finding are two approaches commonly used to accomplish this. *Screening* typically involves investigating large numbers of people, either in a widespread population-based effort or in a more focused fashion within a specific target population. In the primary care setting, screening often translates to investigation of largely asymptomatic patients. This can be costly and time-consuming, often with a low return on effort. *Case-finding* is a more targeted approach, relying on identifying patients with a higher probability of having the disease in question. In primary care diagnosis of respiratory disease, case-finding with the use of symptoms and risk factors is a more realistic approach. We focused on a case-finding approach that begins with a patient's presentation of symptoms and signs suggestive of respiratory disease.

Although lists of symptoms and risk factors are frequently listed as diagnostic aids, specific question items are infrequently tested for their ability to discriminate persons with and without disease. We searched the literature for questionnaire instruments demonstrated

to be valid for diagnosis. In cases where no validated instrument was available, we searched for individual question items, or created and tested new items for this purpose. In all cases, we sought instruments that could be easily administered in a primary care setting with a minimum administrative burden. Since many of the instruments described in the literature were developed for epidemiological or other scientific investigation, they can be quite lengthy, and yet there was little evidence to suggest that a longer instrument improved diagnostic ability. In order to facilitate easy use, we aimed for relatively short question lists, with a target of 5–10 items.

The case-finding tools adopted for the integrated IPCRG guidelines are described below.

- The Allergic Rhinitis Questionnaire was developed by a working group of specialists based primarily upon expert opinion [35].
- The Childhood Asthma Questionnaire was developed based on literature review, a Delphi approach, and a synthesis of question items previously tested and shown to be predictive [36–39].
- The Adult Asthma Questionnaire was developed from a synthesis of question items previously tested and shown to be predictive [40–43].
- The COPD Questionnaire was developed based on literature review, retrospective data analyses, and a Delphi approach. Using prospective methodology, this set of questions has been validated and shown to support diagnosis in a primary care setting [44].
- The Differential Diagnosis Questionnaire was developed along similar lines to the COPD Questionnaire. Using a similar validation process, this tool has been demonstrated to support diagnosis in a primary care setting [45].

Once a patient is placed in a likely diagnostic category using the appropriate questionnaire, more specific guidance can be derived from the summary documents provided by IPCRG, or by referring to the more detailed evidence-based guidelines from ARIA [1], GINA [2], and GOLD [3]. These provide guidance on interpretation of history, physical examination and response to treatment over time, as well as the results of diagnostic tests when available.

Conclusions

Diagnosis of the patient with chronic respiratory disease in the primary care setting requires an integrated approach based upon patients' symptomatic presentation. The recommended

IPCRG approach to diagnosis of respiratory diseases in the primary care setting utilizes a combination of triage by age, initial diagnostic assessment using patient self-reported information (questionnaires and data collection forms), and different levels of confirming evidence based upon available infrastructure.

- For patients with respiratory symptoms, a self-administered questionnaire or data collection form is used based upon age and risk factors.
- This can be easily and efficiently implemented in the practice setting and will provide initial diagnostic assessment that can then be confirmed by the physician.
- Under minimal infrastructure conditions, the initial assessment is confirmed based on findings from history, physical examination, and response to treatment over time.
- Under optimal infrastructure conditions, more definitive testing using spirometry with reversibility testing, peak flow measurement, and allergic testing may be added as needed to provide additional confirmation.
- Patients for whom initial diagnostic assessments cannot be confirmed in the context of available infrastructure should be strongly considered for specialist evaluation.

We believe that this provides a unifying approach to the diagnosis of respiratory diseases which integrates major guideline recommendations and which can be implemented in any primary care setting.

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