

# Pediatria



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#### REVIEW ARTICLE

### Chronic obstructive pulmonary diseases in children\*



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#### **KEYWORDS**

Child; Chronic obstructive pulmonary disease; Chronic cough

#### **Abstract**

*Objectives*: To verify and describe the main events related to the diagnosis and management of chronic obstructive pulmonary diseases in children (COPDC) and adolescents, considering the interrelated physiopathology, genetic, and environmental characteristics.

Sources: Relevant literature from PubMed was selected and reviewed.

Summary of the findings: COPDC have an environmental and/or genetic origin and its manifestation has manifold genotypes, phenotypes, and endotypes. Although COPDC has no cure, it can be clinically controlled. Chronic cough is the main symptom and bronchiectasis can be present in several COPDC patients. The management of COPDC is more effective if based on guidelines and when treatment regimen adherence is promoted. Oral and inhaled corticosteroids, bronchodilators, inhaled antibiotics, and treatment of pulmonary exacerbation (PE) are the bases of COPDC management, and should be individualized for each patient.

Conclusions: Correct diagnosis and knowledge of risk factors and comorbidities are essential in COPDC management. Procedures and drugs used should be based on specific guidelines for each COPDC case. Treatment adherence is critical to obtain the benefits of management. COPDC clinical control must be evaluated by the decrease in PEs, improved quality of life, reduction of pulmonary function loss, and lung structural damage. For most cases of COPDC, monitoring by interdisciplinary teams in specialized reference centers with surveillance strategies and continuous care leads to better outcomes, which must be evaluated by decreasing pulmonary function damage and deterioration, better prognosis, better quality life, and increased life expectancy.

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#### PALAVRAS-CHAVE

Criança; Doença pulmonar obstrutiva crônica; Tosse crônica

#### Doenças pulmonares obstrutivas crônicas na criança

#### Resumo

Objetivos: Verificar e descrever os principais eventos relacionados ao diagnóstico e manejo das doenças pulmonares obstrutivas crônicas em crianças (DPOCC) e adolescentes, tendo em vista a fisiopatologia, características genéticas e ambientais inter-relacionadas.

Fonte dos dados: Revisão na base de dado PUBMED com seleção de referências relevantes. Síntese dos dados: As DPOCC têm origem ambiental e/ou genética e se manifestam com diversos genótipos, fenótipos e endótipos e, embora possam ser controladas, não têm cura. O principal sintoma é a tosse crônica e muitas cursam com bronquiectasia. O manejo tem maior eficácia se baseado em guidelines e se a adesão ao regime terapêutico for estimulada e comprovada. Corticóides orais e inalatórios, broncodilatadores, antibióticos inalados e tratamento das exacerbações pulmonares (EP) são vigas mestras do manejo e devem ser individualizados para cada DPOCC.

Conclusões: Nas DPOCC é fundamental o diagnóstico correto, conhecer os fatores de risco e comorbidades. Os procedimentos e os medicamentos devem ser baseados em *guidelines* específicos para cada DPOCC. Adesão ao tratamento é fundamental para obter os benefícios do manejo. O controle deve ser avaliado pela diminuição das EP, melhora na qualidade de vida e redução da evolução da perda da função e dano estrutural pulmonar. Para a maioria das DPOCC, o acompanhamento por equipes interdisciplinares em centros de referência especializados, com estratégias de vigilância e acolhimento contínuos, conduz a melhores desfechos que devem ser avaliados pela diminuição da deterioração do dano e da função pulmonar, melhor prognóstico, melhor qualidade de vida e aumento da expectativa de vida.

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#### Introduction

Chronic obstructive pulmonary disease (COPD) is typically shown in the literature as evidence, in most cases, of the damage caused by smoking in adults older than 40 years. Without any direct and exclusive association with active long-term smoking, several chronic obstructive pulmonary diseases in children (COPDC) and adolescents progress with deterioration in lung structure and function, causing persistent (fixed) or intermittent (temporary) obstruction to pulmonary flow, secondary to genetic and/or environmental changes that cause airway inflammation and/or infection. Although the symptoms of COPDC are very similar, they have variable etiology, morbidity, physiopathology, prevalence, prognosis, genotypes, and phenotypes. 1,2

While some COPDC have been the object of many studies, such as asthma, cystic fibrosis (CF), recurrent wheezing in infants (RWI), and bronchopulmonary dysplasia (BPD), others are known as "orphan diseases," such as primary ciliary dyskinesia (PCD), non-cystic fibrosis bronchiectasis (NCFB), plastic bronchitis (PB), and bronchiolitis obliterans (BO).

COPDC are characterized by high prevalence of asthma, RWI, and BPD, or low prevalence of BO, CF, PB, PCD, and NCFB. They are noncommunicable diseases, of long duration and slow progression, showing episodes of pulmonary exacerbation (PE), acute or permanent airflow limitation, and significant quality of life impairment.<sup>3</sup> In all, the main pulmonary symptom is chronic cough, reflecting the presence of alterations in the airways, as there are no cough receptors in the alveoli. Another characteristic is the presence of bronchiectasis in many of them.<sup>4</sup>

Most of them, including some of the several clinical forms of asthma, course with neutrophilic airway inflammation,

which contributes to progressive worsening of pulmonary damage and function by releasing: (i) elastase: cleaves elastin and causes bronchiectasis, decreases opsonization and phagocytosis, increases secretion, decrease mucociliary clearance; (ii) DNA: increases the viscosity of secretions; (iii) hydrogen peroxide and other oxidants: causes tissue damage and inactivates  $\alpha$ -1-antitrypsin; (iv) IL-8 and LTB4: attract more neutrophils.<sup>5</sup>

Two aspects of COPDC have been thoroughly studied: the genetic component and environmental aggressions that initiate or exacerbate the diseases. Most COPDC show different genotypes, phenotypes, endotypes, and degrees of severity, require different types of management, and have no cure.<sup>6</sup>

Pneumonia, BPD, BO, and/or RWI in the first years of life constitute risk groups for COPD in the long term and should receive medical follow-up and interventions to prevent the potential impact on long-term respiratory sequelae.<sup>7-9</sup>

In all COPDC, PE is often triggered by viral and/or bacterial infections, pollution, and aeroallergens. The PE manifests as acute respiratory failure of varying intensity, both in asthma and RWI, and as increased cough and chronic infection in CF, BPD, NCFB, and PCD. The signs and symptoms of PE are more frequent and intense at nighttime.

While the majority of acute respiratory diseases can be diagnosed easily and efficiently through history and physical examination, those with a chronic nature may require sophisticated laboratory tests. Continuous and scheduled evaluations by interdisciplinary health teams in specialized centers are required for effective management, better prognosis, and improved quality of life in COPDC.

Establishing protocols based on systematic reviews, meta-analyses, and guidelines allows for gaining control of

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