Financial/nonfinancial disclosures: The author has reported to *CHEST* the following conflicts of interest: Dr Chapman has received compensation for consulting with Boehringer Ingelheim GmbH; CSL Behring; GlaxoSmithKline; Merck Frosst; Novartis AG; Takeda Pharmaceuticals International GmbH; Pfizer, Inc; Hoffman-La Roche, Inc; Merck & Co; and Grifols SA; has undertaken research funded by AstraZeneca; Boehringer Ingelheim GmbH; CSL Behring; Forest Laboratories, Inc; GlaxoSmithKline; Novartis AG; ParanGenix, Inc; Hoffman-La Roche, Inc; Takeda Pharmaceuticals International GmbH; and Grifols SA; and has participated in continuing medical education activities sponsored in whole or in part by AstraZeneca; Boehringer Ingelheim GmbH; GlaxoSmithKline; Grifols SA; Merck Frosst; Novartis AG; Takeda Pharmaceuticals International GmbH; and Pfizer, Inc. Dr Chapman holds the GSK-CIHR Research Chair in Respiratory Health Care Delivery at the University Health Network.
Correspondence to: Kenneth R. Chapman, MD, FCCP, Asthma & Airway Centre, University Health Network, Toronto Western Hospital, Room 7-451 E Wing, 399 Bathurst St, Toronto, ON, M5T 2S8, Canada; e-mail: kchapman@ca.inter.net

DOI: 10.1378/chest.12-0981

References

1. Rodrigo GJ, Neffen H. Comparison of indacaterol with tiotropium or twice-daily long-acting β-agonists for stable COPD: a systematic review. *Chest*. 2012;142(5):1104-1110.
2. Chowdhury BA, Seymour SM, Michele TM, Durmowicz AG, Liu D, Rosebraugh CJ. The risks and benefits of indacaterol—the FDA's review. *N Engl J Med*. 2011;365(24):2247-2249.
3. Nelson HS, Weiss ST, Bleecker ER, Yancey SW, Dorinsky PM; SMART Study Group. The Salmeterol Multicenter Asthma Research Trial: a comparison of usual pharmacotherapy for asthma or usual pharmacotherapy plus salmeterol. *Chest*. 2006;129(1):15-26.
4. O'Donnell TV. Asthma mortality—Maori and nonMaori. *N Z Med J*. 1987;100(836):722.
5. Beasley R, D'Souza W, Te Karu H, et al. Trial of an asthma action plan in the Maori community of the Wairarapa. *N Z Med J*. 1993;106(961):336-338.
6. Garrett JE, Mulder J, Wong-Toi H. Reasons for racial differences in A & E attendance rates for asthma. *N Z Med J*. 1989;102(864):121-124.
7. Canadian Institute for Health Information. *Canadian Lung Association, Health Canada, Statistics Canada. Respiratory Disease in Canada*. Ottawa, ON, Canada: Health Canada; 2001.
8. Stoloff SW, Stempel DA, Meyer J, Stanford RH, Carranza Rosenzweig JR. Improved refill persistence with fluticasone propionate and salmeterol in a single inhaler compared with other controller therapies. *J Allergy Clin Immunol*. 2004;113(2):245-251.
9. Calverley PM, Anderson JA, Celli B, et al; TORCH investigators. Salmeterol and fluticasone propionate and survival in chronic obstructive pulmonary disease. *N Engl J Med*. 2007;356(8):775-789.
10. Gershon A, Croxford R, To T, et al. Comparison of inhaled long-acting β-agonist and anticholinergic effectiveness in older patients with chronic obstructive pulmonary disease: a cohort study. *Ann Intern Med*. 2011;154(9):583-592.

Improving Asthma Management

Time for an Action Plan

Asthma self-management education has been an essential component of the National Asthma Education and Prevention Program guidelines since their initial publication in 1991.[1] Although asthma self-management education made sense intuitively, there was limited evidence of benefit at first. Subsequent research has shown improved health outcomes and a reduction in asthma-related health-care costs with self-management programs in patients with chronic asthma.[2] As a result, updated National Asthma Education and Prevention Program asthma guidelines in 1997 and 2007 gave this educational component greater emphasis.[3,4] Recommended as part of such programs are (1) asthma information and self-management training, (2) self-monitoring with symptoms or peak expiratory flow measurements, (3) regular clinician assessment, and (4) a written action plan. Evidence-based assessment of these items individually has demonstrated the greatest benefit from asthma education and self-monitoring. A limited number of studies have assessed the effect of a written action plan separate from self-management programs in general. A Cochrane meta-analysis of these studies concluded they provided insufficient evidence that action plans as the sole intervention improve asthma outcomes.[5]

In this issue of *CHEST* (see page 1143), Patel et al[6] evaluate written asthma action plans in the context of behaviors associated with asthma control and patient satisfaction with care. The authors conducted a cross-sectional analysis of baseline data collected as part of a National Institutes of Health trial evaluating an asthma self-management program in 808 women with asthma so as to determine specifically the benefit of a written asthma action plan. Asthma control was self-reported and was not a primary outcome. Fewer than one-half of the women in the study had received an action plan. Women given a written asthma action plan, when compared with women without such a plan, were significantly more likely to take their asthma medications as prescribed (84% vs 73%), own a peak flow meter (85% vs 57%), initiate discussion about asthma with a physician (64% vs 46%), and be satisfied with their asthma control (89% vs 79%). The frequency of symptoms and asthma control were not different between the two groups. After adjusting for asthma control, annual household income, and medical specialty of asthma care, patients without an asthma action plan had a doubling of the adjusted OR for dissatisfaction with their asthma care compared with patients with such a plan. The authors conclude that lack of an asthma action plan could adversely affect health-care outcomes by decreasing behaviors associated with optimal asthma control.

The current study suggests that use of a written plan for asthma self-management is associated with asthma medication adherence and patient satisfaction with asthma control and care. One wishes the study

had shown that asthma control was improved by provision of an action plan, but this was not an intended outcome of the study and the study design precluded definitive assessment of this. There was no control for the effects of physician intervention, which might have influenced several measured parameters. In addition, asthma symptoms and medication use were self-reported and were not verified by medical or pharmacy record review. The specific components of the written action plans given to patients were not determined. This may be important, because written action plans incorporating two to four action points and both inhaled and oral corticosteroids for treatment of exacerbations improved asthma health outcomes over those without these elements.[7] The single-center nature of the study and the homogeneous population limit the generalizability of the findings.

Despite these limitations, the study by Patel et al[6] addresses some important issues in the care of patients with asthma, a disease whose prevalence is especially high in adult women.[8] They found that use of written asthma action plans was associated with positive asthma self-management behaviors and correlated with patient satisfaction with medical care. A recent study by Ducharme et al[9] showed that children with asthma exacerbations who were given a written action plan in the ED were more likely to fill their prescription for prednisone and have well-controlled asthma in follow-up 28 days later, compared with those not given such a plan. In the present study, patients provided an action plan were more likely to initiate discussion about asthma with their physician. Such discussions may be useful, as shown by Wilson et al[10] in a study of adults with asthma in which a negotiated treatment plan that accommodated patient goals and preferences led to improved medication adherence and clinical asthma outcomes.

While we await studies that provide definitive evidence of the benefit of written asthma action plans alone, substantial evidence supports the use of such plans as part of asthma self-management programs.[2,4] Nonetheless, current clinical practice often does not meet the standards set by national guidelines. In a study of patients with acute asthma in an ED, only 26% of adults had received a written action plan prior to presentation.[11] Only 34.2% of people with asthma in the United States interviewed in the National Health Interview Survey reported being given a written asthma action plan. Indeed, only 48% of patients in the well-educated population in the current study had received such a plan. How can we make the provision of action plans a part of our regular clinical practice? Here, information and communication technology may come to our aid. Asthma action plans can be downloaded from the NHLBI website (www.nhlbi.nih.gov/health/public/lung/asthma/asthma_actplan.pdf). Asthma action plans can also be developed for incorporation into electronic medical records to be given to the patient on discharge from the hospital, ED, and outpatient clinic. This requires forethought and planning but once incorporated into practice can become routine. Guidelines suggest targeting high-risk populations, such as patients with moderate or severe asthma, a history of frequent exacerbations, or poorly controlled asthma.[4] Telemanagement may improve our ability to provide updated action plans in response to patients' self-monitoring.[12] The data now exist to support the incorporation of written asthma action plans into clinical practice as part of an asthma education program. The study by Patel et al[6] suggests this may improve patient medication adherence and satisfaction with both asthma control and medical care.

Mary E. Strek, MD, FCCP
Chicago, IL

Affiliations: From the University of Chicago, Department of Medicine, Section of Pulmonary and Critical Care.
Financial/nonfinancial disclosures: The author has reported to *CHEST* the following conflicts of interest: Dr Strek conducts clinical research trials for the University of Chicago. None of the trials is related to the topic of this editorial.
Correspondence to: Mary E. Strek, MD, FCCP, the University of Chicago, 5841 S Maryland Ave, MC6076, Chicago, IL 60637; e-mail: mstrek@medicine.bsd.uchicago.edu

DOI: 10.1378/chest.12-1115

References

1. National Asthma Education and Prevention Program. *Expert Panel Report: Guidelines for the Diagnosis and Management of Asthma.* Bethesda, MD: National Institutes of Health; 1991. NIH publication 1-3642.
2. Gibson PG, Powell H, Wilson A, et al. Self-management education and regular practitioner review for adults with asthma. *Cochrane Database Syst Rev*. 2002(3):CD001117.
3. National Asthma Education and Prevention Program. *Expert Panel Report 2: Guidelines for the Diagnosis and Management of Asthma.* National Heart, Lung and Blood Institute website. http://www.nhlbi.nih.gov/guidelines/asthma. Accessed April 11, 2012.
4. National Asthma Education and Prevention Program. *Expert Panel Report 3: Guidelines for the Diagnosis and Management of Asthma.* National Heart, Lung and Blood Institute website. http://www.nhlbi.nih.gov/guidelines/asthma. Accessed April 11, 2012.
5. Toelle BG, Ram FSF. Written individualised management plans for asthma in children and adults. *Cochrane Database Syst Rev*. 2004(2):CD002171.
6. Patel MR, Valerio MA, Sanders G, Thomas LJ, Clark NM. Asthma action plans and patient satisfaction among women with asthma. *Chest*. 2012;142(5):1143-1149.
7. Gibson PG, Powell H. Written action plans for asthma: an evidence-based review of the key components. *Thorax*. 2004; 59(2):94-99.
8. Centers for Disease Control and Prevention (CDC). Vital signs: asthma prevalence, disease characteristics, and self-management education: United States, 2001-2009. *MMWR Morb Mortal Wkly Rep*. 2011;60(17):547-552.

9. Ducharme FM, Zemek RL, Chalut D, et al. Written action plan in pediatric emergency room improves asthma prescribing, adherence, and control. *Am J Respir Crit Care Med*. 2011; 183(2):195-203.
10. Wilson SR, Strub P, Buist AS, et al; Better Outcomes of Asthma Treatment (BOAT) Study Group. Shared treatment decision making improves adherence and outcomes in poorly controlled asthma. *Am J Respir Crit Care Med*. 2010;181(6): 566-577.
11. Camargo CA Jr, Reed CR, Ginde AA, Clark S, Emond SD, Radeos MS. A prospective multicenter study of written action plans among emergency department patients with acute asthma. *J Asthma*. 2008;45(7):532-538.
12. McLean S, Chandler D, Nurmatov U, et al. Telehealthcare for asthma. *Cochrane Database Syst Rev*. 2010(10):CD007717.

Out-of-Proportion Pulmonary Hypertension

A Paradigm for Rare Diseases

Out-of-proportion pulmonary hypertension (PH) is defined as an unjustified degree of PH that occurs in patients suffering from different types of parenchymal lung diseases (ie, COPD, idiopathic pulmonary fibrosis [IPF], and so forth). The concept of out-of-proportion PH was introduced quite recently in the field of PH. Until a few years ago, the term cor pulmonale was used to indicate the development of PH related to parenchymal diseases and exposure to chronic hypoxia, leading to chronic respiratory failure and, consequently, to right-sided heart failure. However, in recent years, a better understanding of the mechanisms underlying the structural remodeling of the pulmonary vascular bed has raised doubts as to whether everything can be explained by the presence of cor pulmonale. In fact, some patients may develop extremely high pulmonary artery pressure that cannot be explained solely by hypoxia and rearrangement of the pulmonary vascular bed. Due to a growing use of right-sided heart catheterization to select appropriate candidates for lung transplantation, a vast category of patients suffering from parenchymal lung diseases (often accompanied by minor pulmonary impairment on pulmonary function test and/or CT scan) with an unexplained severe degree of PH was brought to the attention of physicians. In these patients, the development of moderate to severe PH, which cannot be explained by the degree of parenchymal lung disease and hypoxia, has been termed "out-of-proportion" PH, and an arbitrary value of >35 mm Hg mean pulmonary artery pressure has been selected to identify this category of patients.

In the field of pulmonary rare diseases, this condition was first described in a series of patients with advanced pulmonary Langerhans cell hystiocytosis (PLCH) and was subsequently confirmed by several studies.[1,2] In PLCH, several mechanisms have been proposed to play a role in the development of an out-of-proportion PH. These mechanisms include pulmonary vasculitis involving arterioles and venules, vascular remodeling similar to that observed in patients with idiopathic pulmonary arterial hypertension (iPAH), and inflammation, a phenomenon commonly observed in patients with different types of PH and recently outlined also in patients with iPAH.[3] The report from Le Pavec and coworkers,[4] which is published in this issue of *CHEST* (see page 1150), adds something more to our knowledge. This retrospective analysis evaluated 29 patients with PLCH-associated PH followed for many years in two experienced centers. Thanks to the results obtained, we now know that an isolated decline in the diffusion capacity of the lung for carbon monoxide could be indicative of the development of PH in patients with PLCH and should lead to more accurate investigations; moreover, we know that drugs for iPAH can be useful to treat patients with PLCH who develop moderate to severe PH. In Le Pavec and colleagues'[4] study, 14 patients received iPAH therapies after baseline evaluation. Most patients received initial therapy with an endothelin receptor antagonist ($n = 10$) or phosphodiesterase-5 inhibitor ($n = 5$, two of whom received a combination therapy), and only one patient was treated with inhaled iloprost. Five patients (35%) ultimately received a second PAH agent, either instead of or in combination with the initial drug. All patients had a significant improvement in hemodynamics, and such improvement persisted over time.

Other diseases can also affect the prevalence and severity of PH. One of these is lymphangioleiomyomatosis. This disease shares with PLCH a cystic radiologic pattern; however, PH rarely occurs in patients with lymphangioleiomyomatosis and, if present, is much less severe than that observed in PLCH.[5] In other granulomatous diseases such as sarcoidosis, several mechanisms participate in the development of PH and are not related to the severity of pulmonary function test impairment. As elegantly described by Nunes and colleagues,[6] these mechanisms include sarcoid granulomatous vasculitis, mechanical compression of the large pulmonary artery by adenopathies, and distortion of the vascular bed. However, patients with IPF and PH probably represent the more intriguing area of investigation. In patients with IPF, the normal vascular response to chronic hypoxia consists of medial hypertrophy and intimal fibrosis of pulmonary arterioles; the pathologic manifestations in patients with IPF and out-of proportion PH are still not well known, and the prevalence of out-of proportion PH is still a matter of debate.[7-10] We also know from Cottin and colleagues' studies[11,12] that severe PH occurs more frequently in a particular category of patients suffering from