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REVIEW

Hypersensitivity pneumonitis

Dildar DUMAN(ID) | Clinic of Pulmonology, Memorial Göztepe Hospital, İstanbul, Türkiye

ABSTRACT

Hypersensitivity pneumonitis

Hypersensitivity pneumonitis (HP) is an immune-mediated interstitial lung disease that develops in susceptible individuals following exposure to identifiable or unidentified antigens. Traditionally, HP was classified into acute, subacute, and chronic forms. However, the 2020 ATS/JRS/ALAT guideline introduced a new dichotomous classification into non-fibrotic and fibrotic HP, emphasizing the presence or absence of fibrosis as the key determinant based on radiologic and histopathologic findings. The 2021 CHEST expert panel statement further refined the diagnostic algorithm by integrating exposure history, high-resolution computed tomography patterns, and bronchoalveolar lavage lymphocytosis into a probability-based diagnostic framework. Most recently, the 2025 German S2k guideline proposed an extended three-tiered system—acute, chronic non-fibrotic, and chronic fibrotic HP—and provided updated recommendations for diagnostic pathways and antifibrotictherapy. In light of these evolving perspectives, this review aims to comprehensively discuss the diagnostic and therapeutic strategies for HP, comparing and contrasting the approaches and key recommendations across majör international guidelines.

Key words: *Hypersensitivity pneumonitis; interstitial lung disease; fibrosis*

ÖZ

Hipersensitivite pnömonisi

Hipersensitivite pnömonisi (HP), tanımlanabilen ya da tanımlanamayan antijenlere maruziyet sonrası duyarlanmış bireylerde gelişen immün aracılı bir interstisyel akciğer hastalığıdır. Geleneksel olarak HP, akut, subakut ve kronik biçimlerde sınıflandırılmaktaydı. Ancak 2020 ATS/JRS/ALAT kılavuzu, radyolojik ve/veya histopatolojik olarak fibrozisin varlığına veya yokluğuna dayalı olarak non-fibrotik ve fibrotik HP şeklinde ikili bir sınıflama getirmiştir. 2021 CHEST uzman paneli bildirisi ise, antijen maruziyet öyküsü, yüksek rezolüsyonlu bilgisayarlı tomografi paternleri ve bronkoalveoler lavaj lenfositozunu içeren olasılığa dayalı tanısal algoritma yaklaşımını benimseyerek tanı sürecini daha sistematik hale getirmiştir. En güncel olarak 2025 Alman S2k kılavuzu, HP'yi akut, kronik non-fibrotik ve kronik fibrotik olmak üzere üç basamakta sınıflandırmayı önermiş ve tanı ile tedavi (özellikle antifibrotik tedavi) yaklaşımlarında önemli güncellemeler sunmuştur. Bu derlemede, HP'nin tanısal ve tedaviye yönelik stratejileri, başlıca uluslararası kılavuzların ortak ve farklı yönleri karşılaştırmalı biçimde ele alınarak güncel literatür ışığında tartışılacaktır.

Anahtar kelimeler: *Hipersensitivite pnömonisi; interstisyel akciğer hastalığı; fibrozis*

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Address for Correspondence

Dr. Dildar DUMAN
Clinic of Pulmonology,
Memorial Göztepe Hospital
İSTANBUL-TÜRKİYE
e-mail: dildaryetis@yahoo.com

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Data Sharing Statement: The data that support the findings of this study are available from the corresponding author upon reasonable request.

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INTRODUCTION

Hypersensitivity pneumonitis (HP) is an immune-mediated interstitial lung disease (ILD) that develops in sensitized individuals following exposure to identified or unidentified antigens. Historically, HP was referred to as extrinsic allergic alveolitis. Traditionally, HP was classified into acute, subacute, and chronic forms; however, the 2020 ATS/JRS/ALAT guideline redefined the disease into fibrotic and non-fibrotic categories based on the radiologic and/or histopathologic presence of fibrosis (1,2). The 2021 CHEST expert panel statement further refined the diagnostic framework by integrating exposure assessment, high-resolution computed tomography (HRCT) patterns, bronchoalveolar lavage (BAL) lymphocytosis, and histopathology into a structured probability-based approach. Most recently, the 2025 German S2k guideline proposed a three-tier classification—acute, chronic non-fibrotic, and chronic fibrotic HP—reflecting the dynamic continuum from inflammation to fibrosis (3) (Figure 1). In addition to these evolving classification systems, all three guidelines have introduced updates in diagnostic algorithms and therapeutic recommendations, including the role of antifibrotic agents in progressive fibrotic phenotypes. This review will discuss HP's diagnostic and therapeutic strategies in light of the latest international guidelines and evidence-based updates.

Definition and epidemiology

HP is an immune-mediated ILD occurring in sensitized individuals after exposure to recognized or unrecognized antigens. Also known as extrinsic allergic alveolitis, HP exhibits significant variability in clinical presentation and disease course, depending on the type of inhaled antigen, exposure intensity, duration, and immunologic response (1-3,4). Due to diagnostic difficulties, providing accurate incidence or prevalence figures is challenging. The prevalence of HP may vary based on geographic location, cultural practices, and even seasonal factors (1,3-5). The reported incidence ranges between 0.3 and 0.9 per 100.000 individuals (1). Among farmers, the prevalence of "farmer's lung" ranges from 1% to 19%, and among pigeon breeders, HP prevalence varies between 6% and 20%. The low incidence rates may be attributed to underdiagnosis (1,3,5,6). HP affects men and women at nearly equal rates and can occur at any age, although it is most commonly diagnosed in adults aged 50-60 years (5). The literature indicates that HP including farmer's lung, is more prevalent in non-smokers than in smokers (7-9). HP is the third most common type of ILD, following idiopathic pulmonary fibrosis (IPF) and connective tissue disease-associated ILD, comprising approximately 15% of all ILD cases (10,11).

Pathophysiology

HP is a non-IgE-mediated hypersensitivity reaction involving the lung parenchyma and small airways, where genetic and environmental factors contribute. The disease is complex, as only a small proportion of individuals exposed to potential antigens develop HP. Even among sensitized individuals, some may remain asymptomatic, while others do not develop sensitization at all, suggesting a genetic predisposition (1,3,10).

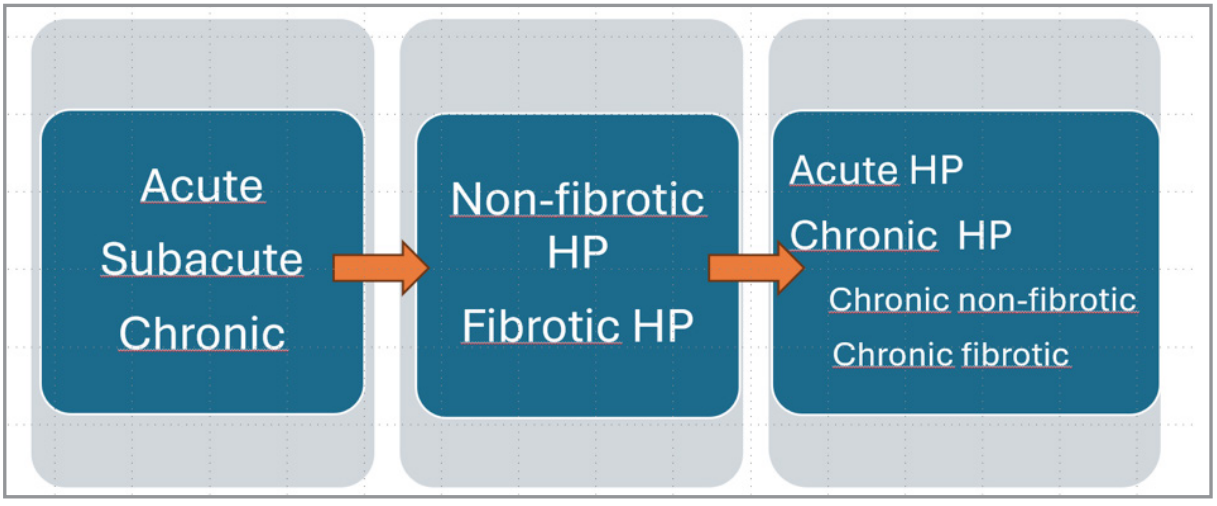


Figure 1. The evolution of the classification of hypersensitivity pneumonitis.

Genetic predisposition has also been described in chronic HP, where MUC5B rs35705950 and short telomere length are associated with greater fibrosis extent, histopathological features of usual interstitial pneumonia (UIP), and reduced survival, suggesting shared pathobiology with IPF (12,13).

The detection of high titers of antigen-specific IgG antibodies in the serum of HP patients points to a role for humoral immunity, whereas the presence of T-lymphocytic alveolitis indicates cellular immune mechanisms (1,3,10). In the early phase of bird-related HP, the immune response is characterized by a mixed Th1/Th2 profile. With disease progression, although Th1 activity persists, cytokine patterns increasingly indicate a shift toward a Th2/Th17-dominated response (14).

A detailed history regarding antigen exposure must be obtained. Identifying the causative antigen is crucial for diagnosis and for implementing preventive strategies. Detailed exposure history followed by laboratory testing (e.g., precipitating IgG antibodies) can be utilized (1-3,15). Structured questionnaires assessing environmental, regional and occupational exposures can be helpful. As occupational exposures play a central role in HP, a structured occupational history should be an integral part of the clinical evaluation of symptomatic patients (16). A positive specific antibody test supports the diagnosis when the clinical picture is compatible, but a negative result does not exclude HP. According to the German HP guidelines, standardized questionnaires and specific IgG antibody testing are recommended for all newly diagnosed ILD patients suspected of HP (3). Inhalation challenge tests, associated with the risk of severe reactions, should not be performed outside specialized centers and are not routinely recommended by any guidelines (1-5). Despite extensive evaluations, a causative antigen cannot be identified in up to 50% of patients.

Over 200 antigens have been implicated in HP, including bacteria, fungi, animal proteins, plant proteins, chemicals, and metals (Table 1). Bird proteins and molds are the most commonly reported antigens (1-5).

Diagnostic approach

The primary goal in the diagnostic approach to HP is to establish an accurate diagnosis using the least invasive methods available while maintaining high diagnostic reliability (3).

The diagnosis of HP relies on three key pillars:

- Evidence of antigen exposure (based on clinical history),
- Evidence of sensitization (positive specific IgG antibodies and/or BAL lymphocytosis),
- Evidence of ILD compatible with HP (based on HRCT findings and/or lung biopsy) (3).

Radiology

If an interstitial pattern is suspected on chest radiography, the patient should be evaluated with HRCT. Notably, chest radiographs can appear normal in approximately 20% of HP cases (3).

For the diagnosis of HP, HRCT should be performed using thin slices (<1.5 mm), during deep inspiration, volumetrically, and without contrast. Expiratory imaging is recommended to assess air trapping (17,18).

Typical radiologic features of HP include mid-to-upper zone predominance, ground-glass opacities, centrilobular nodules, air trapping, mosaic attenuation, and

Table 1. Common antigens in hypersensitivity pneumonitis and their sources*

Antigen	Source	Associated Disease
Avian proteins	Bird feathers, droppings, serum proteins (e.g., pigeon, parakeet, canary)	Bird Fancier's lung
Thermophilic *Actinomycetes*	Moldy hay, straw, grain, silage (e.g., *Saccharopolyspora rectivirgula*, *Thermoactinomyces vulgaris*)	Farmer's lung
Aspergillus and *Penicillium* species	Moldy cheese, compost, moldy wood (e.g., *Aspergillus fumigatus*, *Penicillium casei*)	Cheese washer's lung, compost lung
Mycobacterium avium complex	Contaminated hot tub water, warm water therapy pools	Hot tub lung
Isocyanates	Paints, sprays, polyurethane foams (e.g., toluene diisocyanate, methylene diphenyl diisocyanate)	Chemical worker's lung

the "three-density pattern". Ground-glass opacities suggest alveolitis, centrilobular micronodules point to small airway disease, and mosaic attenuation areas reflect bronchiolar involvement (1-3,17,18). The presence of ground-glass opacities interspersed with normal lung and air trapping areas constitutes the characteristic "three-density pattern" for chronic HP (18) (Figure 2).

In fibrotic HP, fibrosis tends to be randomly distributed, often with relative sparing of the lower lobes, distinguishing it from IPF and non-specific interstitial pneumonia (NSIP), where lower lobe dominance is typical (17).

Bronchoalveolar lavage

In newly diagnosed ILD patients with suspected HP, BAL with differential cell count analysis is recommended to increase diagnostic confidence. BAL lymphocytosis (>30%) strongly supports the diagnosis of HP and helps distinguish it from other ILDs such as sarcoidosis. In fibrotic HP, lymphocytosis tends to be more modest and may even fall within normal ranges, particularly in smokers or elderly patients. Nevertheless, persistent BALF lymphocytosis, even in fibrotic disease, remains diagnostically useful in differentiating fibrotic HP from IPF. The CD4/CD8 ratio, however, should not be used as a diagnostic criterion. Beyond cytological evaluation, BAL is also valuable for excluding infectious etiologies, including *Mycobacterium* species (1,3,10,19).

Histopathology

If a lung biopsy is required to diagnose acute or chronic nonfibrotic HP, TBLB or TBLC should be considered. For chronic fibrotic HP, TBLC is the preferred approach if a biopsy is needed. However, if a prior TBLC did not provide meaningful findings or if TBLC is not feasible, a surgical lung biopsy should be performed (3).

The histopathology of nonfibrotic HP typically includes cellular interstitial pneumonia with a bronchiolocentric pattern, cellular bronchiolitis with organizing pneumonia and foam cells, and poorly formed, non-necrotizing granulomas mainly in the peribronchiolar interstitium (3,20,21). Fibrotic HP is defined by interstitial fibrosis, often with granulomas. Key features include centrilobular, bronchiolocentric, or bridging fibrosis, where fibrosis extends between centrilobular and perilobular regions (21). Additional findings may include giant cell reactions with cholesterol clefts or oxalate crystals, and occasional areas of acute injury. Fibrotic HP can show honeycombing similar to IPF, but unlike IPF, it can also involve the upper lobes and often appears asymmetrically (3,21).

In the most recent interstitial pneumonia classification, the airway-centered pattern previously associated with HP has been incorporated under the term bronchiolocentric interstitial pneumonia. This terminology aims to distinguish morphologic patterns from multidisciplinary clinical diagnoses. Accordingly, cases with an airway-centered interstitial pneumonia pattern but without an identifiable

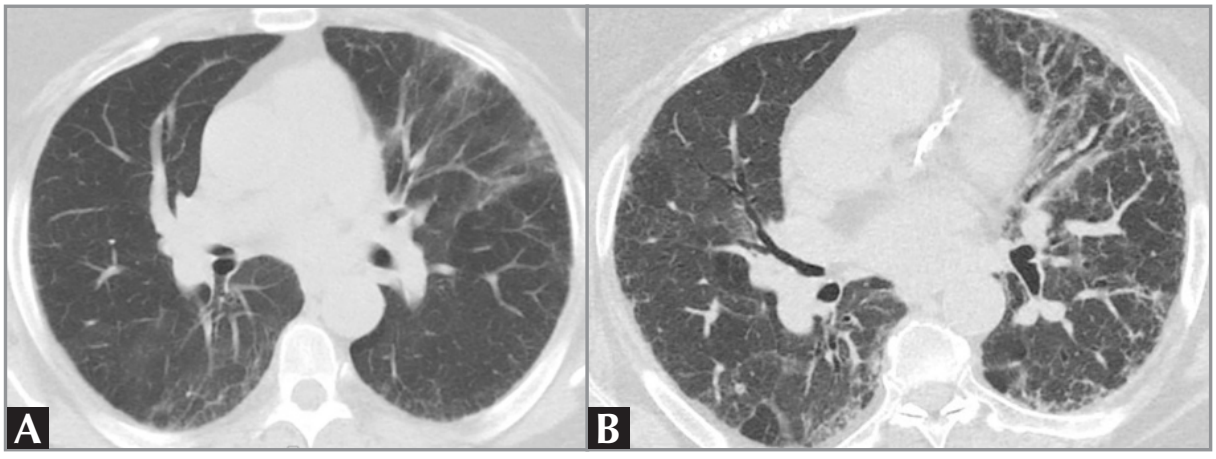


Figure 2. HRCT images of the same patient taken six years apart. **(A).** Acute HP: Bilateral ground glass areas. **(B).** Chronic fibrotic HP. Radiology: Traction bronchiectasis in the left lingular area, ground glass areas involving both central and peripheral areas of the lung. Subpleural reticulations. Three density signs typical for chronic HP consisting of air trapping area, normal lung area, and ground glass area in the right lower lobe.

antigen exposure are now referred to as idiopathic BIP. This conceptual update refines the classification of HP-related and non-HP airway-centered ILDs, emphasizing the importance of multidisciplinary evaluation and comprehensive exposure assessment in establishing a definitive diagnosis (22).

Diagnostic criteria for acute ad chronic HP were shown as below:

Diagnostic criteria for acute HP

Primary diagnostic criteria (All five must be met):

1. Documented exposure to a potential antigen source,
2. Recurrent symptomatic episodes occurring 4 to 8 hours post-exposure,
3. Elevated specific IgG antibody titers against the suspected antigen,
4. Inspiratory crackles detected on clinical examination,
5. HRCT findings consistent with inflammatory HP.

If all five criteria are satisfied, the diagnosis of acute HP is confirmed.

Alternative invasive diagnostic criteria (if all primary criteria are not met):

- BAL lymphocytosis >30%,
- Histopathological findings typical or probable for acute or non-fibrotic HP on lung biopsy,
- Positive results from inhalation provocation, re-exposure, or antigen avoidance tests.

Diagnostic criteria for chronic fibrotic HP

Major diagnostic criteria

(Diagnosis is confirmed if at least three of the following four criteria are met):

1. Documented exposure to a potential antigen source,
2. Elevated specific IgG antibody titers against a potential antigen and/or BAL lymphocytosis (>30%),
3. HRCT findings consistent with fibrotic HP,
4. Histopathological findings typical or probable for chronic fibrotic HP on lung biopsy.

Differences between non-fibrotic and fibrotic actinomycetes HP were summarized in Table 2.

Treatment

Treatment of acute HP

The cornerstone of treatment for acute HP is identifying and completely eliminating the causative antigen. If the offending antigen can be identified and exposure can be terminated, and if the patient exhibits only minimal clinical symptoms, minimal functional impairment, and non-fibrotic radiologic findings (such as mild ground-glass opacities and centrilobular nodules), treatment may consist solely of antigen avoidance (e.g., removing pet birds) (3,23).

Table 2. Differences between non-fibrotic and fibrotic hypersensitivity pneumonitis

Feature	Non-Fibrotic (Inflammatory) HP	Chronic Fibrotic HP
Definition	Reversible inflammation predominant	Irreversible fibrosis predominant
Clinical course	Subacute or acute onset, symptoms develop rapidly	Chronic, insidious onset, progressive shortness of breath
Symptoms	Cough, fever, chills, fatigue, shortness of breath	Long-standing cough, exertional dyspnea
HRCT findings	Ground-glass opacities, mosaic perfusion, air trapping	Reticular infiltrates, honeycombing, traction bronchiectasis
BAL findings	Lymphocytosis (>30%)	Lymphocytosis may be present, but fibrosis is dominant
Biopsy findings	Inflammatory cell infiltration, granuloma-like structures	Fibrosis, honeycomb structure, chronic changes
Specific IgG	High	High
Treatment	Elimination of antigen, short-term steroids	Antigen removal, immunosuppressants, antifibrotics
Prognosis	Generally good, improves with treatment	Worse, may be progressive

HP: Hypersensitivity pneumonitis, HRCT: High-resolution computed tomography patterns, BAL: Bronchoalveolar lavage.

However, if the patient's symptoms—such as cough and dyspnea—progress significantly over days to weeks, or if radiologic findings show more extensive ground-glass opacities and centrilobular nodules, or if there is notable functional decline, short-term corticosteroid therapy should be considered. Although randomized controlled trials are lacking, the generally recommended initial regimen mirrors that used in sarcoidosis, consisting of 0.5 mg/kg/day of prednisone (or equivalent) or a 40 mg/day fixed dose. The dose should be tapered gradually over 2-3 months, aiming for eventual discontinuation (3,23).

Treatment of chronic HP

As in acute HP, antigen avoidance remains the first and most crucial step in the management of chronic HP. When there is evidence of an inflammatory component (e.g., centrilobular nodules or ground-glass opacities on HRCT, BAL lymphocytosis, NSIP-like histology, or the presence of non-necrotizing granulomas), anti-inflammatory treatment is indicated (3,24).

Initial treatment typically involves corticosteroids. If necessary, corticosteroid-sparing immunosuppressive agents such as azathioprine or mycophenolate mofetil may be added (3,9). The recommended initial maximum dose of prednisolone is 0.5 mg/kg/day. If the patient responds, a gradual taper should be implemented over 2-3 months, targeting a maintenance dose of less than 10 mg/day, maintained for at least six months (3,24).

Treatment with prednisone, either as monotherapy or in combination with azathioprine, has been shown to confer clinical benefit in both fibrotic and non-fibrotic HP patients. Favorable predictors of therapeutic response included fever following antigen exposure, a BAL lymphocyte fraction exceeding 54%, an RV/TLC ratio >120% predicted, and the presence of centrilobular nodules on HRCT (25).

Antifibrotic therapy

In cases where significant but non-dominant fibrotic features are present, a combination of anti-inflammatory and antifibrotic therapies may be considered either simultaneously or in close succession (3,26).

In patients with chronic HP who meet the criteria for progressive pulmonary fibrosis (PPF), antifibrotic therapy becomes indicated (26-29). Evidence suggests that purely anti-inflammatory treatment may not be effective and may even negatively impact prognosis in patients with predominant fibrosis (3,24). In patients with short telomere lengths, immunosuppression has been associated with worse outcomes and increased mortality; thus, in such cases, initiating antifibrotic therapy without prior immunosuppression can be recommended (30).

The INBUILD trial demonstrated that chronic HP constituted the largest subgroup (26%) of progressive fibrosing ILD patients, and that treatment with nintedanib significantly reduced the annual decline in forced vital capacity (FVC) and slowed disease progression (31). The 2022 ATS/ERS clinical practice guideline for IPF/PPF recommended nintedanib for patients with PPF, noting that more research is needed for pirfenidone in this context (27). Although some studies support the potential benefit of pirfenidone in PPF and fibrotic HP, pirfenidone has not yet been officially approved for these indications (32,33).

Lung transplantation

Lung transplantation should be considered if disease progression persists despite optimal medical therapy. Post-transplant survival rates in patients with HP are typically superior to those observed in individuals with IPF (3). In patients who develop hypoxemia, pulmonary rehabilitation should not be overlooked, alongside oxygen therapy, to strengthen respiratory muscles and improve quality of life (27,33). Ideally, all treatment decisions should be made by a multidisciplinary IAH council (3).

Prognosis

Survival varies depending on the form of HP: In non-fibrotic HP, survival rates are excellent. In fibrotic HP, the median survival is approximately 9.2 years (24).

Several factors have been identified as predictors of reduced survival, including persistent and high-intensity antigen exposure, diminished functional capacity [reflected by decreased FVC and diffusing capacity for carbon monoxide (DLCO)], GAP score ≥4 points, exercise-induced hypoxemia, radiological findings of extensive fibrosis, absence of lymphocytosis in BAL, histopathological patterns consistent with UIP or fibrotic NSIP, presence of pulmonary hypertension, genetically shortened telomere length, and the MUC5B promoter polymorphism (3,34,35). Adjusted for age, fibrosis, FVC%, DLCO%, and smoking history, survival was longer in patients with identified inciting antigen exposure than in those with unidentified exposure (8.7 vs. 4.8 years) (36).

CONCLUSION

The 2025 German S2k guideline introduces a refined classification as acute, chronic non-fibrotic, and chronic fibrotic HP. It also emphasizes the radiologic categorization of HRCT findings as either inflammatory or fibrotic HP. Additionally, separate diagnostic criteria have been established for acute or non-fibrotic HP and chronic fibrotic HP, allowing for more precise diagnosis and management.The guideline further provides differentiated recommendations for anti-inflammatory and/or antifibrotic therapy in chronic fibrotic HP, reflecting the distinct pathophysiology and treatment needs of these subgroups.

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CONFLICT of INTEREST

The author declare that they have no conflict of interest.

AUTHORSHIP CONTRIBUTIONS

Concept/Design: DD

Analysis/Interpretation: DD

Data Acquisition: DD

Writing: DD

Clinical Revision: DD

Final Approval: DD

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