Comment on: Successful management of pulmonary hypertension with baricitinib in a dermatomyositis patient

Sirs,

We read with great interest the case by Rizzo *et al*., reporting the successful use of baricitinib in a patient with dermatomyositis (DM)-associated pulmonary hypertension (PH) (1). PH is a haemodynamic condition defined by a mean pulmonary arterial pressure >20 mmHg at rest, confirmed via right heart catheterisation (2). PH is classified into five groups, based on differing pathophysiological mechanisms, clinical characteristics, and therapeutic approaches. Importantly, only Group 1 PH should be labelled as pulmonary arterial hypertension (PAH). In the article by Rizzo *et al*., the term "PAH" appears to be used interchangeably for Group 1 and Group 3 PH.

Group 1 PH/PAH involves increased pulmonary vascular resistance due to structural or functional alterations in the pulmonary arteries (3), possibly caused by hereditary factors, drug/toxin exposure, and underlying conditions including connective tissue diseases (CTDs), HIV infection, portal hypertension, congenital heart disease, and schistosomiasis (2, 4). Differently, Group 3 PH arises from chronic lung diseases and/or hypoxia, particularly relevant in CTDs such as systemic sclerosis, where interstitial lung disease (ILD) is common. The remaining PH groups include: Group 2, associated with left heart disease; Group 4, resulting from chronic thromboembolic or other pulmonary artery obstructions; and Group 5, encompassing multifactorial or unclear mechanisms (2). Accurate classification is essential, impacting prognosis and treatment strategies.

Idiopathic inflammatory myopathies (IIM), including DM, are systemic autoimmune rheumatic disorders characterised by chronic skeletal muscle inflammation and progressive muscle weakness. In IIM, PH is most often linked to extensive ILD, corresponding to Group 3 PH, as observed in antisynthetase syndrome (5). Relevant to this case, lung function in IIM-ILD may stabilise or improve with appropriate immunosuppressive therapy (6). Some reports have described possible cases of isolated PAH in patients with IIM, including those with DM. However, the occurrence of this subtype in IIM, as reported by Rizzo *et al*., appears exceedingly rare (3, 7).

The DM patient described underwent a diagnostic evaluation that included pulmonary function tests (normal), a high-resolution chest computed tomography scan (with no evidence of significant interstitial lung disease or cardiac involvement), transthoracic echocardiography (with indirect signs of PH), and right heart catheterisation (the gold standard for definite diagnosis of PH, compatible with this condition). Additionally, in our view, the inclusion of a ventilation/perfusion (V/Q) lung scan could have further enriched the diagnostic assessment by excluding the possibility of chronic thromboembolic pulmonary hypertension (Group 4 PH) (2, 3). This test is currently recommended by the European Society of Cardiology (ESC) and the European Respiratory Society (ERS) as part of the diagnostic work-up in patients with suspected PH (2). This consideration is pertinent for several reasons: baricitinib, the Janus kinase inhibitor (JAKi) administered, is contraindicated in patients with a history or presence of thrombosis (8); DM is associated with a higher risk of venous thromboembolism (9); the use of intravenous immunoglobulin, previously administered to the described patient, can sometimes lead to thromboembolic events (10).

Regarding the therapeutic strategy employed, given the lack of robust evidence for the use of JAKi in DM-associated PH, we consider that other options could have been explored, namely the concomitant use of an endothelin receptor antagonist (ERA) and a phosphodiesterase 5 inhibitor (PDE5i), supported by the 2022 ESC/ERS guidelines (2). Although the clinical presentation of this case suggests improvement, the gold-standard method to confirm the resolution of PH would be reassessment through right heart catheterisation.

In conclusion, we would like to emphasise that diagnosing and characterising pulmonary PH subtypes represent a complex clinical challenge. Accurate classification requires a comprehensive evaluation integrating clinical, imaging, haemodynamic, and laboratory data, and is best conducted within a multidisciplinary setting. A precise and prompt diagnosis is crucial to optimise patient outcomes and ensure the most appropriate, evidence-based management.

C. Pinto Oliveira[1,2], *MD*
M.D. Pascoal[1,2], *MD*
A.R. Prata[1,2], *MD*
D. Carvalho[3], *MD*
M.G. Castro[4], *MD*
E. Dourado[1,2], *MD*

[1]Rheumatology Department, Unidade Local de Saúde Região de Aveiro;
[2]Aveiro Rheumatology Research Centre, Egas Moniz Health Alliance, Aveiro;
[3]Cardiology Department, Unidade Local de Saúde Região de Aveiro;
[4]Heart and Vessels Department, Cardiology, Hospitais da Universidade de Coimbra, Unidade Local de Saúde de Coimbra, Portugal.

Please address correspondence to:
Cláudia Pinto Oliveira
Rheumatology Department,
Unidade Local de Saúde da Região de Aveiro,
Avenida Artur Ravara,
3810-164 Aveiro, Portugal.
E-mail:
claudia.oliveira.71800@ulsra.min-saude.pt

Competing interests: none declared.

References

1. RIZZO C, CAMARDA F, LA BARBERA L, GUGGINO G: Successful management of pulmonary hypertension with baricitinib in a dermatomyositis patient. *Clin Exp Rheumatol* 2025; 43(2): 390. https://doi.org/10.55563/clinexprheumatol/c6an8c
2. HUMBERT M, KOVACS G, HOEPER MM *et al*.: 2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension. *Eur Heart J* 2022; 43(38): 3618-731. https://doi.org/10.1093/eurheartj/ehac237
3. SANGES S, SOBANSKI V, LAMBLIN N *et al*.: Pulmonary hypertension in connective tissue diseases: what every CTD specialist should know - but is afraid to ask! *Rev Med Interne* 2024; 45(1): 26-40. https://doi.org/10.1016/j.revmed.2023.10.450
4. KOVACS G, BARTOLOME S, DENTON CP *et al*.: Definition, classification and diagnosis of pulmonary hypertension. *Eur Respir J* 2024; 64(4): 2401324. https://doi.org/10.1183/13993003.01324-2024
5. HERVIER B, MEYER A, DIEVAL C *et al*.: Pulmonary hypertension in antisynthetase syndrome: prevalence, aetiology and survival. *Eur Respir J* 2013; 42(5): 1271-82. https://doi.org/10.1183/09031936.00156312
6. CORREIA BP, CAMPANILHO-MARQUES R, DOURADO E *et al*.: Myositis-associated interstitial lung disease: the experience of a tertiary center. *J Clin Med* 2024; 13(20): 6085. https://doi.org/10.3390/jcm13206085
7. BHANSING KJ, VONK-NOORDEGRAAF A, OOSTERVEER FP, VAN RIEL PL, VONK MC: Pulmonary arterial hypertension, a novelty in idiopathic inflammatory myopathies: insights and first experiences with vasoactive therapy. *RMD Open* 2017; 3(1): e000331. https://doi.org/10.1136/rmdopen-2016-000331