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# Significant Reduction of Lung Disease on Baricitinib Therapy in a Patient With COPA Syndrome

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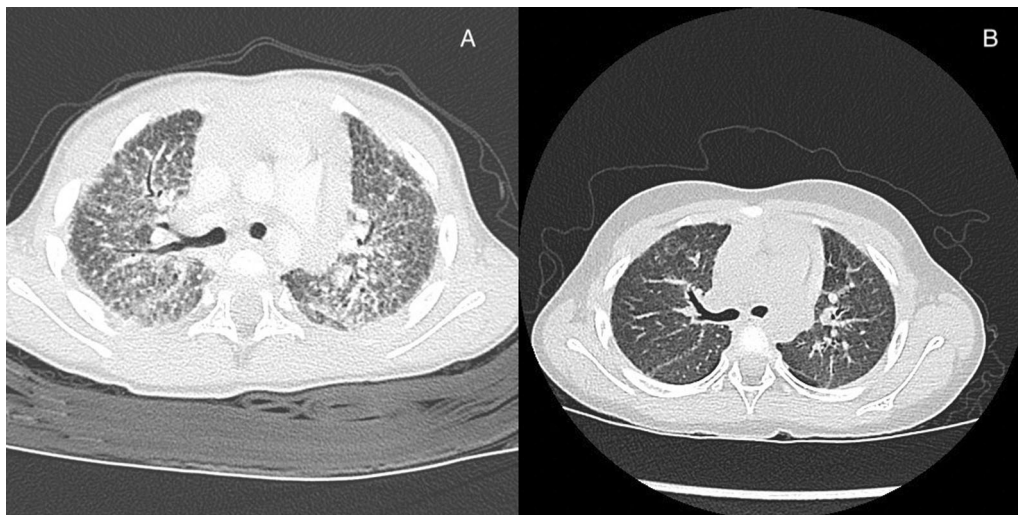
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To the Editor,

A 4-year-old girl affected by COPA syndrome with joint and pulmonary involvement [1] was on treatment with baricitinib, a Janus kinase inhibitor, since the age of 2. The arthritis quickly resolved. While she had radiographic evidence of significant lung disease (Figure 1A), she has always been asymptomatic, without digital clubbing, nail ridging or abnormal oxy-hemoglobin saturation.

After 2 years of baricitinib therapy, a lung CT scan was performed (Figure 1B). The images show a remarkable improvement, with a reduction of pulmonary interstitium thickening and an almost complete disappearance of ground-glass opacities if compared with CT scan underwent at diagnosis (Figure 1A). Concurrently, blood sampling proved a negative result of SIGLEC1 and of the interferon score [2], which was highly positive at diagnosis (interferon score: 24, normal value:  $\leq 2.2$ ).



**FIGURE 1** | Lung CT scan at diagnosis, showing diffuse marked thickening of the pulmonary interstitium with ground glass opacities. (A). Lung CT scan after 2 years, showing remarkable improvement of pulmonary interstitial thickening and significant reduction of previously described ground glass opacities. (B).

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It is well known that JAK inhibitors are very effective in treating arthritis [3]. To the best of our knowledge, this is the first report of significant lung improvement in a patient with COPA disease on baricitinib therapy. This could be due to an early treatment when there could be a physiological margin for recovery.

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### Author Contributions

**Elettra Zuliani:** writing–original draft. **Elisa Canepari:** writing–original draft. **Serena Pastore:** writing–review and editing, formal analysis. **Francesca Neri:** software, data curation. **Alessandra Tesser:** supervision. **Erica Valencic:** supervision. **Giulia Gortani:** supervision. **Massimo Maschio:** supervision. **Alberto Tommasini:** writing–review and editing, supervision, formal analysis. **Andrea Taddio:** writing–review and editing, supervision, formal analysis.

### Ethics Statement

The study was approved by the Internal Review Board of the IRCCS Burlo Garofolo of Trieste with the grant ID RC 34/18. Parents of the child gave consent for publication of this report.

### Conflicts of Interest

The authors declare no conflicts of interest.

### Data Availability Statement

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

### References

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