

Erratum to: Longitudinal, natural history study reveals the disease burden of idiopathic multicentric Castleman disease

Mateo Sarmiento Bustamante,^{1*} Sheila K. Pierson,^{1*} Yue Ren,² Adam Bagg,³ Joshua D. Brandstadter,⁴ Gordan Srkalovic,⁵ Natalie Mango,¹ Daisy Alapat,⁶ Mary Jo Lechowicz,⁷ Hongzhe Li,² Frits van Rhee,⁸ Megan S. Lim⁹ and David C. Fajgenbaum¹

¹Center for Cytokine Storm Treatment & Laboratory, Department of Medicine, Perelman School of Medicine, University of Pennsylvania, Philadelphia, PA; ²Department of Biostatistics, Epidemiology, and Informatics, Perelman School of Medicine, University of Pennsylvania, Philadelphia, PA; ³Department of Pathology and Laboratory Medicine, University of Pennsylvania, Philadelphia, PA; ⁴Division of Hematology/Oncology, Perelman School of Medicine, University of Pennsylvania, Philadelphia, PA; ⁵Sparrow Herbert-Herman Cancer Center, Michigan State University College of Human Medicine, Lansing, MI; ⁶Department of Pathology, College of Medicine, University of Arkansas for Medical Sciences, Little Rock, AR; ⁷Department of Hematology and Medical Oncology, Emory University School of Medicine and Winship Cancer Institute, Atlanta, GA; ⁸Myeloma Center, Winthrop P. Rockefeller Cancer Institute, University of Arkansas for Medical Sciences, Little Rock, AR and ⁹Department of Pathology and Laboratory Medicine, Memorial Sloan Kettering Cancer Center, New York City, NY, USA.

**MSB and SKP contributed equally as first authors.*

The article by Sarmiento Bustamante *et al.*¹ recently published in *Haematologica*, contained severity data for patients with idiopathic multicentric Castleman disease ± 90 days from diagnosis. Upon review, some NOS patients were being counted as having severe disease based on criteria outside of the 90-day window. Additionally, a number of typographical errors were identified. The authors' interpretation of their data and findings are not changed by the corrections, listed below, of these errors.

Point 1

On page 2198 of the article by Sarmiento Bustamante *et al.*¹ in Table 1, the numbers in the Severity at Diagnosis row should read:

iMCD
Severe: 71 (72.4)
Mild/moderate: 27 (27.6)
Data not available: 4

NOS:
Severe: 14 (37.8)
Mild/moderate: 23 (62.2)
Data not available: 4

The numbers in the TAFRO column remain unchanged.

Point 2

On page 2199 of the article by Sarmiento Bustamante *et al.*¹ in the sentence reproduced below, the data on ages should be presented as median (interquartile range) values instead of their mean (standard deviation) values:

Correct sentence:

“TAFRO patients tended to be more commonly male (TAFRO 62% compared to NOS 49%) and younger (median [IQR]: 31.4

[17.9–47.1] vs. 39.3 [31.3–50.0]) than NOS patients.”

Point 3


On page 2199 of the article by Sarmiento Bustamante *et al.*¹ in the sentence reproduced below, 98 patients had sufficient information to assess disease severity at presentation and 71/98 (72.4%) patients presented with severe disease, not 77/100 (77.0%).

Correct sentence:

“Of the 98 patients with sufficient information to determine disease severity at diagnosis, 71 (72.4%) initially presented with severe disease.”

Point 4

On page 2199 of the article by Sarmiento Bustamante *et al.*¹ in the sentences reproduced below and Figure 1A, the *P* value for the trend in severity between age groups is 3.1×10^{-5} , not 5.3×10^{-4} . Additionally, 94.4% of patients <30 and 53.8% of patients between 30 and 60 years old presented with severe disease, not 94.6% and 62.2%, respectively. The legend to Figure 1A should state that 94% of patients under the age of 30 years presented with severe disease at diagnosis, not 95%.

Correspondence: D.C. Fajgenbaum
davidfa@pennmedicine.upenn.edu
Received: August 19, 2024.
Accepted: August 19, 2024.
<https://doi.org/10.3324/haematol.2024.286500>
©2024 Ferrata Storti Foundation
Published under a CC BY-NC license 

Correct sentences:

“We examined severity by age range visually and observed that the majority of patients <30 or >60 years of age presented with severe disease (Figure 1A). When we examined this trend statistically, we found a significant difference among the groups ($P=3.1\times10^{-5}$). Patients <30 and patients >60 years old were more likely to have severe disease (94.4% and 90.0%, respectively) as compared to patients between 30 and 60 years old (53.8%) (Online Supplementary Table S2).”

Point 5

On page 2200 of the article by Sarmiento Bustamante *et al.*,¹ in the sentence reproduced below, 98 patients had sufficient information to assess disease severity at presentation and 71/98 (72.4%) patients presented with severe disease, not 77/102.

Correct sentence:

“Among the 98 patients with sufficient information to determine disease severity at diagnosis, 71 (72.4%) presented with severe disease at diagnosis, including 57 (93.4%) TAFRO

patients and 14 (37.8%) NOS patients (Table 1).”

Point 6

On page 2200 of the article by Sarmiento Bustamante *et al.*,¹ in the legend to Figure 2, in the sentence reproduced below for part (B), TAFRO patients had a median hospitalization time of 36 days, not 35 days, in a 12-month period:

Correct sentence:

“(B) A sample of the USA population from the 2018 National Health Information Survey (NHIS), a proxy for the general population, spent significantly less time hospitalized in a 12-month period (median [IQR]: 0 [0-0], mean [SD]: 0.24 [1.3] days) than either TAFRO patients (median [IQR]: 36 [18-61], mean [SD]: 46.0 [42.0] days; $P<2.2\times10^{-16}$) or NOS patients (median [IQR]: 0 [0-4], mean [SD]: 4.9 [9.0] days; $P=8.3\times10^{-13}$) spent hospitalized in the year around diagnosis.”

Point 7

On page 2201 of the article by Sarmiento Bustamante *et al.*,¹ in the sentence reproduced below, the proportion of patients experiencing cardiovascular symptoms and laboratory ab-

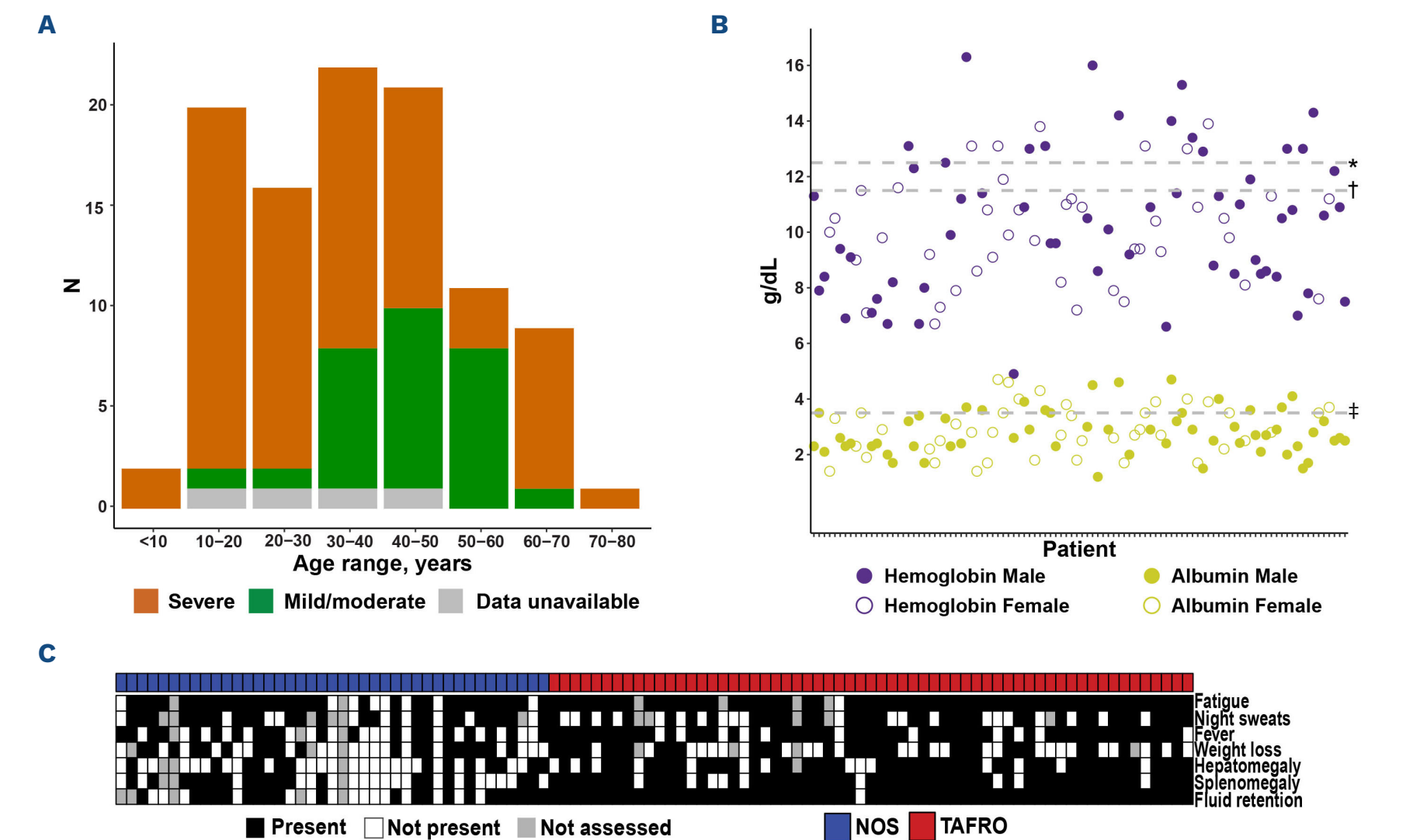


Figure 1. Patients with idiopathic multicentric Castleman disease demonstrate severe disease at diagnosis. (A) A large proportion of patients with idiopathic multicentric Castleman disease (iMCD) across all ages had severe disease. Notably, 94% of patients under the age of 30 years presented with severe disease at diagnosis. (B) Within the cohort of 102 iMCD patients, 87 (85.3%) had anemia and 81 (79.4%) had hypoalbuminemia at diagnosis. *Lower limit of normal for hemoglobin in males (12.5 g/dL). †Lower limit of normal for hemoglobin in females (11.5 g/dL). ‡Lower limit of normal for albumin (3.5 g/dL). (C) A large majority of patients presented with clinical symptoms, which ranged from mild to severe. Patients with both clinical subtypes of iMCD (TAFRO and NOS) demonstrated clinical abnormalities. TAFRO: thrombocytopenia, anasarca, fever/elevated C-reactive protein, reticulin fibrosis/renal failure, and organomegaly; NOS: not otherwise specified.

normalities was 68 (66.7%), not 63 (61.8%):

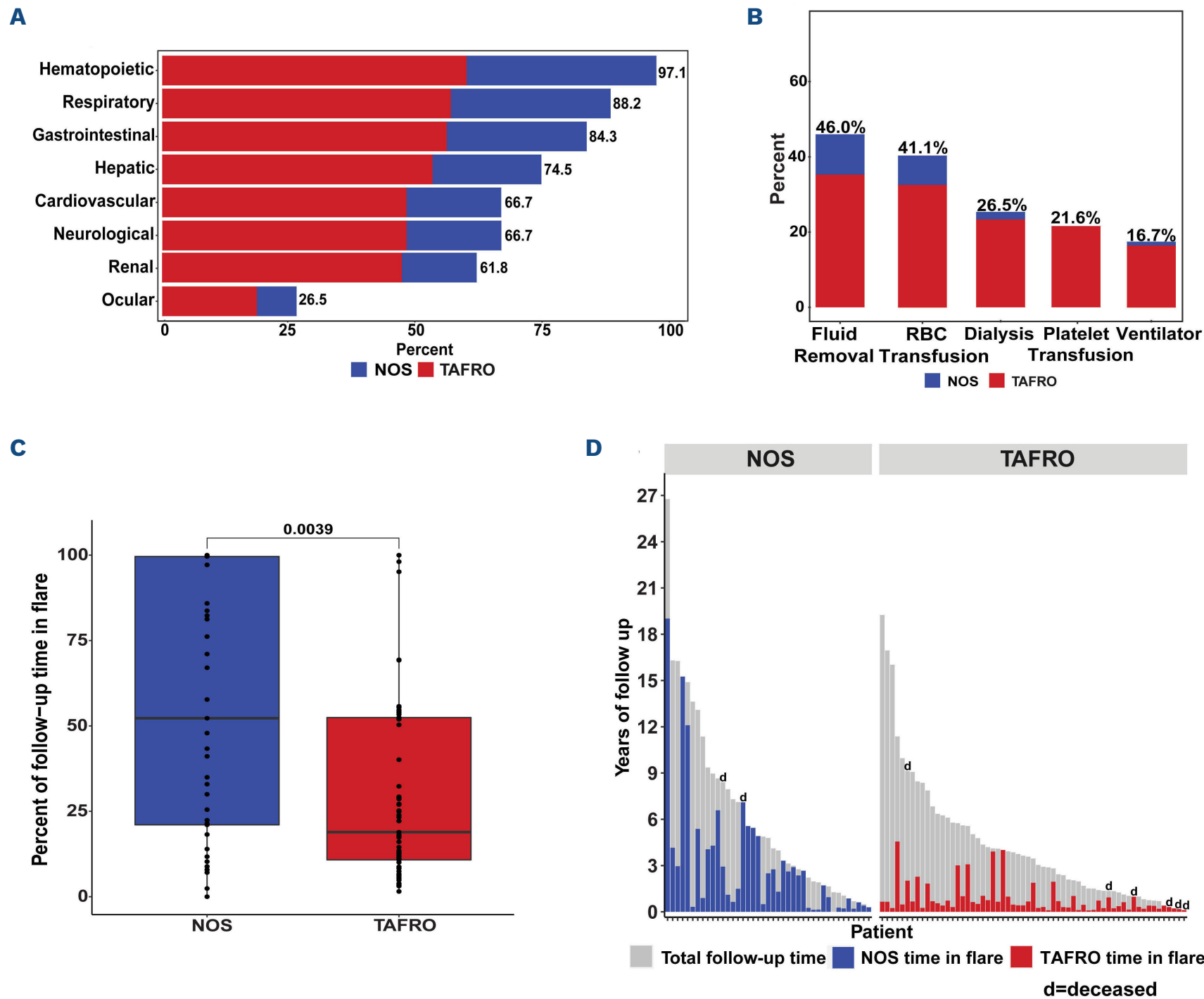
Correct sentence:

“We also found large proportions of patients experiencing symptoms or laboratory abnormalities related to other organ systems, such as respiratory (N=90, 88.2%), gastrointestinal (N=86, 84.3%), hepatic (N=76, 74.5%), neurological (N=68, 66.7%), cardiovascular (N=68, 66.7%), renal (N=63, 61.8%),

and ocular (N=27, 26.5%) systems (Figure 3A, *Online Supplementary Tables S5 and S6*).”

Point 8

On page 2202 of the article by Sarmiento Bustamante *et al.*,¹ in the legend to Figure 3, in the sentence reproduced below for part (B), ventilator and dialysis and their respec-



Corrected Figure 3. Patients with idiopathic multicentric Castleman disease experience a range of organ system involvement, require various hospital interventions, and demonstrate ongoing flares. (A) Organ system involvement in idiopathic multicentric Castleman disease (iMCD) ranked from most frequently observed to least frequently observed. Over 97% of the iMCD cohort experienced some hematopoietic dysfunction. Notably, both patients with TAFRO (thrombocytopenia, anasarca, fever/elevated C-reactive protein, reticulin fibrosis/renal failure, and organomegaly) and those with iMCD not otherwise specified (NOS) experienced significant organ system involvement and dysfunction. (B) The severity of organ dysfunction is reflected by the degree of healthcare intervention(s) required. Over one-quarter of patients (N=27 [26.5%]) required dialysis and 17 (16.7%) patients required the use of a ventilator. Additionally, 47 (46.0%) patients required fluid removal (paracentesis), 42 (41.1%) patients received a red blood cell transfusion, and 22 (21.6%) patients received a platelet transfusion. (C) NOS patients spent a significantly greater proportion of time in flare from presentation until last known information (median [interquartile range]: 52.3% [21.0-99.6]) compared to TAFRO patients (18.9% [10.8-52.5], $W=1673$; $P=0.004$). (D) Each patient is represented by a vertical bar. The bar extends the length of follow-up from the start of the first flare. NOS patients are represented on the left, and TAFRO patients are represented on the right. The blue bars represent the proportion of time NOS patients spent in flare. The red bars represent the proportion of time TAFRO patients spent in flare. The designation ‘d’ indicates a deceased patient. RBC: red blood cells.

tive numbers were flipped:
Correct sentence
“Over one-quarter of patients (N=27 [26.5%]) required dialysis, and 17 (16.7%) patients required the use of a ventilator.”

Point 9
On page 2202 of the article by Sarmiento Bustamante *et al.*¹ in Figure 3A, the neurological involvement was 66.7%, not 48.0% as the published figure indicated, and should therefore have a bar the same length as that for Cardiovascular involvement. *The correct Figure 3 is provided above, together with its corrected legend (see Point 8).*

Point 10
On page 2203 of the article by Sarmiento Bustamante *et al.*¹ in the legend to Figure 4, in the sentence reproduced below for part (B), the number of iMCD patients developing iron deficiency anemia following diagnosis was 14 (13.7%), not 11 (10.8%):
Correct sentence:
“(B) Following the diagnosis of iMCD, patients experienced an array of burdensome comorbidities and morbidities including acute renal failure (N=49, 48.0%), chronic kidney disease/chronic renal insufficiency (N=16, 15.7%) and iron deficiency anemia (N=14, 13.7%) among others.”

Point 11
On page 2204 of the article by Sarmiento Bustamante *et al.*¹

in the sentence reproduced below, 37.8% of NOS patients met criteria for severe disease at presentation, not 51.3%.
Correct sentence:
“Although NOS patients spent significantly less time in hospital, we found that 37.8% met criteria for severe disease at presentation.”

Point 12
On page 3 of the *Online Supplementary Appendix* to the article by Sarmiento Bustamante *et al.*¹ in *Online Supplementary Table S2*, the numbers for patients <30 years old and patients 30-60 years old presenting with severe disease should be:
<30 years (n=36)
Mild/moderate, n (%): 2 (5.6%)
Severe, n (%): 34 (94.4%)
30-60 years (n=52)
Mild/moderate, n (%): 24 (46.2%)
Severe, n (%): 28 (53.8%)
The numbers for patients >60 years remain unchanged.

Point 13
On page 11 of the *Online Supplementary Appendix* to the article by Sarmiento Bustamante *et al.*¹ in *Online Supplementary Table S7*, the numbers for Pneumonia (10 [9.8]) and Chronic Kidney Disease (12 [11.8]) are incorrect and should instead be Pneumonia (11 [10.7]) and Chronic Kidney Disease (16 [15.7]).

References

1. Bustamante MS, Pierson SK, Ren Y, et al. Longitudinal, natural history study reveals the disease burden of idiopathic multicentric Castleman disease. *Haematologica*. 2024;109(7):2196-2206.