Supplementary Appendix

Sarcoidosis Treatment Algorithm Delphi Questionnaire 3 Results

This table contains the questions from the third Delphi questionnaire, the mean and standard deviation of the Likert scale results, information on whether consensus was reached, and all written comments received. Comments have been lightly edited for clarity and anonymity. Bold indicates a consensus for the statement, and a grey background indicates a consensus against.

Question		Mean	Std Dev	Consensus
TREATMENT AT INITIAL PRESENTATION				
Scadding Stage/Chest X-ray				
1.	Patients who are asymptomatic and at Scadding stage 0/1 should usually be treated	-4.68	1.13	Against
2.	Patients who are asymptomatic and at Scadding stage 2/3 should usually be treated	-2.09	2.65	No
3.	Patients who are asymptomatic and at Scadding stage 4 should usually be treated	-1.23	3.05	No
4.	Patients who are symptomatic and at Scadding stage 0/1 should usually be treated	1.09	2.65	No
5.	**Patients who are symptomatic and at Scadding stage 2/3 should usually be treated**	**4.27**	**0.83**	**For**
6.	**Patients who are symptomatic and at Scadding stage 4 should usually be treated**	**3.73**	**1.35**	**For**
7.	Scadding stage / chest X-ray should not be considered in deciding whether to treat a patient	-0.50	2.96	No
8.	**Scadding stage / chest X-ray should be used in conjunction with symptoms, extrapulmonary involvement, and other factors in deciding whether to treat a patient**	**4.68**	**0.48**	**For**
9.	Scadding stage / chest X-ray should be the primary factor in deciding whether to treat a patient	-4.64	0.79	Against
HRCT				
10.	Patients who are asymptomatic with no parenchymal abnormality on HRCT should usually be treated	-4.23	2.20	Against

11.	Patients who are asymptomatic with parenchymal abnormalities but no fibrosis on HRCT should usually be treated	-1.91	3.07	No
12.	Patients who are asymptomatic with fibrosis on HRCT should usually be treated	-0.73	2.85	No
13.	Patients who are symptomatic with no parenchymal abnormality on HRCT should usually be treated	0.09	2.64	No
14.	**Patients who are symptomatic with parenchymal abnormalities but no fibrosis on HRCT should usually be treated**	**3.77**	**1.31**	**For**
15.	**Patients who are symptomatic and with fibrosis on HRCT should usually be treated**	**3.55**	**1.30**	**For**
16.	HRCT should not be considered in deciding whether to treat a patient	-1.77	2.98	No
17.	**HRCT should be used in conjunction with symptoms and other factors in deciding whether to treat a patient**	**4.82**	**0.50**	**For**
18.	HRCT should be the primary factor in deciding whether to treat a patient	-3.41	1.37	Against
19.	**Patients with severe symptoms causing impaired QOL due to pulmonary sarcoidosis should usually be treated**	**4.00**	**1.45**	**For**
Imaging: General				
20.	A radiologic finding should be present before initiating therapy for sarcoidosis	-0.59	3.40	No
FVC				
21.	Patients who are asymptomatic and have FVC >80% should usually be treated	-3.95	2.13	Against
22.	Patients who are asymptomatic and have FVC 70-80% should usually be treated	-1.77	2.49	No
23.	Patients who are asymptomatic and have FVC<70%	0.09	2.99	No
24.	Patients who are symptomatic and have FVC >80% should usually be treated	1.32	2.87	No
25.	**Patients who are symptomatic and have FVC 70-80% should usually be treated**	**3.27**	**1.52**	**For**
26.	**Patients who are symptomatic and have FVC <70%**	**4.27**	**1.12**	**For**
27.	FVC should not be considered in deciding whether to treat a patient	-3.09	2.51	Against
28.	**FVC should be used in conjunction with symptoms and other factors in deciding whether to treat a patient**	**4.86**	**0.35**	**For**
29.	FVC should be the primary factor in deciding whether to treat a patient	-2.59	1.79	Against
DLCO				
30.	Patients who are asymptomatic and have DLCO >80% predicted should usually be treated	-3.82	1.92	Against
31.	Patients who are asymptomatic and have DLCO 70-80% predicted should usually be treated	-2.45	2.60	No
32.	Patients who are asymptomatic and have DLCO <70%	-0.55	2.86	No

33.	Patients who are symptomatic and have DLCO >80% predicted should usually be treated	1.45	2.54	No
34.	**Patients who are symptomatic and have DLCO 70-80% predicted should usually be treated**	**2.77**	**1.97**	**For**
35.	**Patients who are symptomatic and have DLCO <70%**	**3.91**	**1.02**	**For**
36.	DLCO should not be considered in deciding whether to treat a patient	-3.95	0.95	Against
37.	**DLCO should be used in conjunction with symptoms and other factors in deciding whether to treat a patient**	**4.91**	**0.29**	**For**
38.	DLCO should be the primary factor in deciding whether to treat a patient	-3.32	2.17	Against
39.	**Other potential causes of low DLCO should be evaluated before initiating treatment for sarcoidosis based on a low DLCO**	**4.23**	**2.16**	**For**
40.	**Low DLCO values should prompt an investigation of possible pulmonary hypertension**	**4.23**	**1.31**	**For**
FEV1/FVC				
41.	Patients who are asymptomatic and have FEV1/FVC >80% predicted should usually be treated	-3.82	1.65	Against
42.	Patients who are asymptomatic and have FEV1/FVC 70-80% predicted should usually be treated	-2.77	1.88	Against
43.	Patients who are asymptomatic and have FEV1/FVC <80%	-0.82	2.38	No
44.	Patients who are symptomatic and have FEV1/FVC >80% predicted should usually be treated	1.09	2.88	No
45.	Patients who are symptomatic and have FEV1/FVC 70-80% predicted should usually be treated	1.91	2.45	No
46.	**Patients who are symptomatic and have FEV1/FVC <70%**	**3.00**	**1.98**	**For**
47.	FEV1/FVC should not be considered in deciding whether to treat a patient	-2.18	2.11	No
48.	**FEV1/FVC should be used in conjunction with symptoms and other factors in deciding whether to treat a patient**	**4.23**	**1.11**	**For**
49.	FEV1/FVC should be the primary factor in deciding whether to treat a patient	-4.05	1.70	Against
50.	FEV1/FVC is useful as an indicator of obstructive that may require treatment with ICS	2.23	2.47	No
Pulmonary Function Testing: General				
51.	**Changes and trends in pulmonary function tests (FVC, DLCO, FEV1/FVC) are more important than the absolute values**	**4.05**	**1.29**	**For**
Extrapulmonary Disease				
52.	**Assessment of extrapulmonary disease is useful indicator of disease activity**	**3.95**	**1.13**	**For**
53.	**Extrapulmonary disease should be treated if it affects the patient's quality of life**	**4.00**	**1.48**	**For**
54.	**Extrapulmonary disease should be treated if hypercalcemia is present**	**4.73**	**0.88**	**For**

55.	**Asymptomatic patients with evidence of cardiac extrapulmonary disease involvement should usually be treated**	**3.27**	**2.07**	**For**
56.	**Asymptomatic patients with evidence of neurologic extrapulmonary disease involvement should usually be treated**	**3.09**	**1.85**	**For**
57.	**Asymptomatic patients with evidence of ocular extrapulmonary disease involvement should usually be treated**	**3.91**	**1.11**	**For**
58.	**Asymptomatic patients with evidence of renal extrapulmonary disease involvement should usually be treated**	**3.64**	**1.47**	**For**
59.	Asymptomatic patients with evidence of cutaneous extrapulmonary disease involvement should usually be treated	0.18	2.91	No
60.	Asymptomatic patients with evidence of hepatic extrapulmonary disease involvement should usually be treated	-0.86	3.14	No
61.	Please describe any other extrapulmonary disease sites that should prompt initiation of therapy in asymptomatic patients. • Bone • High U-calcium and problems with stones • Central nervous system, cardiac, eye, and skin diseases, and severe bone involvement • Eye or renal involvement, sarcoidosis of the upper respiratory tract • Bone marrow			
62.	**Symptomatic patients with evidence of cardiac extrapulmonary disease involvement should usually be treated**	**4.86**	**0.35**	**For**
63.	**Symptomatic patients with evidence of neurologic extrapulmonary disease involvement should usually be treated**	**4.82**	**0.50**	**For**
64.	**Symptomatic patients with evidence of ocular extrapulmonary disease involvement should usually be treated**	**4.91**	**0.29**	**For**
65.	**Symptomatic patients with evidence of renal extrapulmonary disease involvement should usually be treated**	**4.64**	**0.79**	**For**
66.	**Symptomatic patients with evidence of cutaneous extrapulmonary disease involvement should usually be treated**	**3.73**	**1.24**	**For**
67.	**Symptomatic patients with evidence of hepatic extrapulmonary disease involvement should usually be treated**	**3.32**	**1.84**	**For**

68.	Other extrapulmonary disease sites should prompt initiation of therapy in symptomatic patients	2.50	2.91	No
68.	a. Please list/describe the sites. • Bone • Swollen parotid glands, arthritis • Bone marrow sarcoidosis with low counts, sarcoidosis-associated vitamin D dysregulation with hypercalciuria and recurrent nephrolithiasis of no other cause, gastrointestinal sarcoidosis • Bone involvement. Spinal involvement • Neurologic, eye, liver, or kidney involvement; sarcoidosis of the upper respiratory tract • Bone marrow, spleen			
69.	Extrapulmonary disease should not be considered in deciding whether to treat a patient	-4.27	2.16	Against
70.	**Extrapulmonary disease should be used in conjunction with symptoms and other factors in deciding whether to treat a patient**	**4.55**	**1.30**	**For**
71.	Extrapulmonary disease should be the primary factor in deciding whether to treat a patient	-1.45	2.77	No
Other considerations				
72.	Asymptomatic patients with pulmonary hypertension should usually be treated	0.68	2.78	No
73.	**Symptomatic patients with pulmonary hypertension should usually be treated**	**3.73**	**1.70**	**For**
74.	**Patients with progressive disease should usually be treated**	**4.55**	**0.80**	**For**
75.	How important are each of the following factors as indicators of progressive disease?			
	Worsening symptoms	**4.05**	**1.05**	**For**
	Radiographic decline persistent infiltrate in HRCT	**2.77**	**2.25**	**For**
	Loss of organ function over time (objective evidence of declining PFTs, worsening fibrosis, increasing lesions on spine or heart or eyes	**4.68**	**0.57**	**For**
	New organ involvement	**2.64**	**1.71**	**For**
	Functional status decline and fatigue	**2.77**	**1.66**	**For**
	Worsening blood tests	1.27	2.66	No
	Reduction in 6-minute walk test	2.32	1.76	No
76.	Patients with long-duration, stable should usually be treated	-1.95	2.24	No
77.	How important are each of the following factors in defining long-duration disease?			
	More than 3-6 months with <10% changes in PFT, imaging, or other functional studies	0.73	2.90	No
	Stability from 6 months to >1 year off therapy	2.09	2.69	No

	>1-2 years of stable disease	**3.86**	**1.98**	**For**
78.	How important are each of the following factors in defining stable disease?			
	No progression of symptoms	**3.73**	**1.39**	**For**
	No PFT worsening	**3.95**	**0.90**	**For**
	No new localization or no worsening of known localizations as measured by QOL, CT and PFT	**4.00**	**0.93**	**For**
	Stable function of any organ with sarcoid manifestation over 12 months or more	**4.18**	**1.14**	**For**
79.	Patients with pulmonary sarcoidosis who need oxygen should usually be treated	1.59	2.38	No
80.	The patient's age is an important factor in deciding whether to initiate treatment	-2.00	2.56	No
81.	The patient's comorbidities are an important factor in deciding whether to initiate treatment	2.05	2.70	No
82.	**The patient's ability to comply with prescribed therapy is an important factor in deciding whether to initiate treatment**	**3.05**	**2.06**	**For**
Importance of various factors				
83.	How important are each of the following factors in your decision to treat patients for pulmonary sarcoidosis at presentation?			
	Patient preference	**2.59**	**2.38**	**For**
	Symptom severity and QOL impact	**4.18**	**0.91**	**For**
	Imaging studies	**2.55**	**2.34**	**For**
	Pulmonary function tests	**3.68**	**1.17**	**For**
	Extrapulmonary organ involvement disease	**4.00**	**0.93**	**For**
	Pulmonary hypertension	**3.55**	**2.09**	**For**
	Need for oxygen	**2.86**	**1.93**	**For**
	Progressive disease	**4.68**	**0.57**	**For**
	Stability of disease	**2.64**	**2.57**	**For**
	Long-duration disease	1.55	2.81	No
	PET scan	1.45	2.61	No
	Cardiac MRI	**3.09**	**1.41**	**For**
	Angiotensin converting enzyme (ACE) levels	-1.50	2.72	No
	Lysozyme	-1.59	3.07	No
	Soluble IL-2 levels	-1.18	3.22	No
	Complete blood count/Lymphocyte panel	-0.73	2.86	No

	Echocardiography (echo)	1.86	1.96	No
	Electrocardiography (ECG)	1.95	2.63	No
	Holter monitoring	2.32	2.32	No
	Comprehensive metabolic panel (CMP)	1.23	2.49	No
	Inflammatory markers (ESR and CRP)	-0.55	2.70	No
	Vitamin D and metabolites	0.45	2.39	No
	Urinalysis/urinary calcium	1.50	2.37	No
	Ophthalmologic examination	**3.41**	**2.15**	**For**
	Liver function tests	1.00	2.89	No
83.a. Other (please describe here and rate below): • Bone disease • Neopterin • HLA • Concomitant MAI (*mycobacterium avium intracellulare*) infection and or other bacterial infections				
	83.b Rate the 'Other factor' described above	**2.83**	**2.04**	**For**
Indications for Immediate Treatment				
84.	Treatment should be started immediately for patients with dyspnoea with minimal or no exertion or hypoxemia to reduce the risk of near-term respiratory failure	1.73	3.59	No
85.	**Treatment should be started immediately for patients with severely impaired pulmonary function tests (low DLCO, FVC, or FEV1) to reduce the risk of near-term respiratory failure**	**3.55**	**1.06**	**For**
86.	**Treatment should be started immediately for patients with rapid decreases in pulmonary function tests (low DLCO, FVC, or FEV1) to reduce the risk of near-term respiratory failure**	**4.32**	**0.65**	**For**
87.	**Treatment should be started immediately for patients who require oxygen to reduce the risk of near-term respiratory failure**	**3.23**	**1.34**	**For**
88.	Treatment should be started immediately for extrapulmonary disease in the following sites:			
	Neurological symptoms severe acute onset neurologic disease with neurologic impairment	**4.86**	**0.47**	**For**
	Cardiac arrhythmias or other cardiac symptoms severe acute onset cardiac disease with complete heart block, VT	**4.86**	**0.47**	**For**
	Acute renal failure due to hypercalcemia and/or renal involvement	**4.86**	**0.35**	**For**
	Significant ocular inflammation	**4.86**	**0.35**	**For**

Other organ failure due to sarcoidosis	**4.32**	**1.21**	**For**
Additional Studies			
89. A PET study should be ordered at initial presentation	-2.59	3.11	No
90. An angiotensin converting enzyme (ACE) test should be ordered at initial presentation	-0.68	3.55	No
91. A lymphocyte panel should be ordered at initial presentation	0.27	3.72	No
92. An echocardiogram (echo) should be ordered at initial presentation	1.14	2.68	No
93. An electrocardiogram (ECG) should be ordered at initial presentation	**4.05**	**1.36**	**For**
94. Measurement of total lung capacity (TLC) should be ordered at initial presentation	2.27	3.55	No
95. A 6-minute walk test (6MWT) should be ordered at initial presentation	1.32	2.95	No

96. Other tests/procedures that should be ordered at initial presentation:
    - Serum calcium/24 hr urinary calcium; fungal and Tb studies as indicated by circumstances
    - CT scan
    - HLA typing
    - Fatigue assessment
    - FVC; FEV1; DLCO; pulmonary imaging; ophthalmologic examination
    - Endocrine blood tests
    - Liver enzymes, renal blood tests, calcium
    - ESR, CRP to subsequently evaluate response to treatment
    - FEV1, DLCO, blood count, Kidney/liver function, calcium
    - CXR or HRCT
    - CMP CBC with diff
    - sIL-2
    - Brain MRI in neurosarcoidosis
    - PET
    - Calcium level, 1,25 Vitamin D level
    - HbA1c, bone density
    - CMP, CBC, ocular exam
    - LFTs, Cr, Ca

Selecting Initial Therapy			
Note: only prednisone and repository corticotrophin injection have received FDA approval for the treatment of pulmonary sarcoidosis.			
Corticosteroids			
97. Initial treatment for pulmonary sarcoidosis should usually be oral prednisone unless contraindicated	**3.14**	**2.32**	**For**
98. The usual starting prednisone dose should be 20 mg/day to 40 mg/day	**4.05**	**0.95**	**For**
99. The usual starting prednisone dose should be 40 mg/day	-0.45	2.76	No
100. The usual starting prednisone dose should be 30 mg/day	0.32	2.73	No
101. The usual starting prednisone dose should be 25 mg/day	0.64	2.44	No
102. The usual starting prednisone dose should be 20 mg/day	1.50	2.46	No
103. The usual starting prednisone dose should be 10 mg/day	-1.55	2.46	No
104. The usual starting prednisone dose should be 0.5 mg/kg/day to 0.6 mg/kg/day	-0.36	3.05	No
105. The usual starting prednisone dose should be 0.6 mg/kg/day	-0.50	2.76	No
106. The usual starting prednisone dose should be 0.5 mg/kg/day	0.05	2.72	No
107. The usual starting prednisone dose should be 0.3 mg/kg/day	-0.09	2.56	No
108. The starting dose should be increased in the following conditions:			
Severe symptoms	1.09	2.84	No
Progressive disease	1.64	2.61	No
Extensive disease	1.55	2.99	No
Extrapulmonary involvement	1.68	2.32	No
109. The starting dose should be decreased in the following conditions:			
Diabetes	**3.00**	**2.02**	**For**
Psychosis	**3.68**	**2.01**	**For**
Osteoporosis	**2.55**	**1.97**	**For**
Obesity	2.23	2.07	No
Adjunctive and steroid-sparing therapy			
110. For patients with mild disease, initial treatment should be a steroid-sparing therapy	-1.27	2.71	No
111. Mild disease can be defined as mild to no symptoms and mild to no impairment of lung function with no significant neurologic, cardiac, or ocular findings	**3.55**	**1.47**	**For**

112. If the above definition is not appropriate, how do you define mild disease in this context? Please enter your definition. • Non-progressive over 3-6 month follow up • No renal symptoms • Mild pulmonary disease needs a more objective definition, e.g. dyspnoea score, functional capacity etc, to standardize the definition				
113. Adjunctive steroid-sparing therapy should be started in the following conditions:				
Simultaneously with steroids	0.77	2.60	No	
If a long-duration of steroid therapy is expected (e.g., for chronic disease)	**3.05**	**2.24**	**For**	
If disease progression occurs despite steroid therapy	**4.50**	**0.67**	**For**	
If long-term, low-dose maintenance therapy does not control the disease	**4.45**	**0.80**	**For**	
If extrapulmonary disease is present	**2.55**	**1.95**	**For**	
If there is high risk for steroid-related adverse events	**4.23**	**0.92**	**For**	
If steroid toxicities develop	**4.68**	**0.57**	**For**	
If steroid failure occurs	**4.59**	**0.73**	**For**	
Inhaled corticosteroids				
114. Patients should be treated with inhaled corticosteroids (ICS) in the following indications:				
For symptomatic relief of cough	**3.45**	**1.22**	**For**	
For relief of asthma signs/symptoms (wheezing, dyspnoea, obstructive PFT)	**3.77**	**1.11**	**For**	
As monotherapy in mild disease with obstructive spirometry	2.23	2.45	No	
As a steroid-sparing adjunct to oral steroids	-0.32	2.90	No	
Other (please describe below)	0.23	1.80	No	
Comments: • There are no other indications for ICS • No other conditions • Associated asthma • If air trapping on CT • The steroid sparing should be initiated directly before the side effect occurs • Asthmatic sarcoidosis				
115. If ICS are used, they should be discontinued in the following conditions:				
Toxicity	**4.41**	**0.85**	**For**	

	Failure to stabilize disease	**3.23**	**2.71**	**For**
	Failure to discontinue oral steroids	0.59	3.14	No
	Failure to alleviate cough	**2.55**	**2.42**	**For**
Follow-up				
116.	During the first year of therapy, patients should be seen at least every month	-1.95	2.89	No
117.	During the first year of therapy, patients should be seen at least every 1-3 months	1.27	2.88	No
118.	During the first year of therapy, patients should be seen at least every 3-6 months	1.91	2.69	No
Adjusting Therapy				
119.	If a patient has improved at follow-up, treatment should usually be adjusted by:			
	Withdrawing the steroid	-1.55	3.22	No
	Decreasing the steroid dose (to find the lowest dose that provides satisfactory symptom relief and disease control)	**4.18**	**0.85**	**For**
	No adjustment is needed	-3.86	1.52	Against
	Increasing the steroid dose	-4.68	0.78	Against
	Adding/increasing adjunctive therapy	-2.27	2.39	No
120.	If a patient is stable at follow-up, treatment should usually be adjusted by:			
	Withdrawing the steroid	-2.82	2.52	Against
	Decreasing the steroid dose (to find the lowest dose that provides satisfactory symptom relief and disease control)	**3.36**	**1.84**	**For**
	No adjustment is needed	-1.64	2.79	No
	Increasing the steroid dose	-2.14	3.01	No
	Adding/increasing adjunctive therapy	-0.36	2.89	No
121.	If a patient is stable but experiencing steroid toxicities at follow-up, treatment should usually be adjusted by:			
	Withdrawing the steroid	0.91	3.02	No
	Switching to another agent	2.50	2.67	No
	Trying a drug holiday	-1.05	2.68	No
	Decreasing the steroid dose	**4.00**	**1.07**	**For**
	No adjustment is needed	-4.45	1.14	Against
	Increasing the steroid dose	-4.32	2.25	Against

Adding/increasing adjunctive therapy	**3.64**	**1.56**	**For**
122. If a patient is stable but you are concerned about steroid toxicities at follow-up, treatment should usually be adjusted by:			
Withdrawing the steroid	0.00	3.15	No
Switching to another agent	1.64	2.56	No
Trying a drug holiday	-0.91	2.79	No
Decreasing the steroid dose	**4.00**	**1.23**	**For**
No adjustment is needed	-3.64	2.13	Against
Increasing the steroid dose	-4.68	0.95	Against
Adding/increasing adjunctive therapy	**3.23**	**1.66**	**For**
123. If a patient is stable but he/she is concerned about steroid toxicities at follow-up, treatment should usually be adjusted by:			
Discussion to reassure the patient	**3.00**	**2.73**	**For**
Withdrawing the steroid	-0.82	2.92	No
Switching to another agent	1.41	2.75	No
Trying a drug holiday	-1.00	3.07	No
Decreasing the steroid dose	**3.00**	**2.05**	**For**
No adjustment is needed	-2.18	2.26	No
Increasing the steroid dose	-4.55	0.96	Against
Adding/increasing adjunctive therapy	**2.73**	**1.45**	**For**
124. If a patient is worse, treatment should usually be adjusted by:			
Withdrawing the steroid	-2.50	2.65	No
Decreasing the steroid dose	-2.32	2.53	No
Re-evaluate the diagnosis and treatment	**4.23**	**1.38**	**For**
No adjustment is needed	-4.14	1.25	Against
Increasing the steroid dose	1.14	2.61	No
Adding/increasing adjunctive therapy	**4.14**	**1.08**	**For**
Weaning			
125. Reduction or discontinuation in therapy should be considered for:			
Typical steroid toxicities	**4.27**	**1.55**	**For**

	Excessive cost	-0.32	3.27	No
	Loss of response	**3.32**	**1.99**	**For**
	Disease progression	0.95	3.17	No
	Lack of efficacy	**3.41**	**1.99**	**For**
	Patient intolerance	**4.14**	**0.83**	**For**
	Patient preference (with full understanding of risks and benefits)	**3.50**	**1.06**	**For**
	All patients	2.23	2.37	No
126.	Therapy should be regarded as ineffective if the patient has not improved after:			
	1-4 weeks	-2.32	2.83	No
	1-3 months	0.77	3.26	No
	3-6 months	**3.73**	**1.55**	**For**
127.	Weaning from therapy should be considered for:			
	Toxicity/tolerability issues	**4.50**	**0.80**	**For**
	Treatment failure/lack of efficacy	**4.14**	**1.25**	**For**
	Treatment success/ improvement or resolution of symptoms	**3.91**	**1.15**	**For**
128.	When weaning from steroids is appropriate, weaning should usually be done by slowly tapering the dose over 6-12 months with careful disease monitoring	2.36	3.14	No
129.	When weaning from agents other than steroids, weaning should usually be done by simply stopping the agent	-0.23	3.02	No
130.	Patients can be weaned from steroids and other agents simultaneously	-1.23	3.05	No
Non-biologic agents				
131.	Non-biologic agents should be considered if:			
	A steroid-sparing regimen is needed	**4.27**	**0.88**	**For**
	Steroid toxicities develop	**4.36**	**0.90**	**For**
	Prolonged and/or high-dose steroid use is expected	**4.55**	**0.60**	**For**
	Steroids are not effective	**4.27**	**0.94**	**For**
	Non-biologics should be used in most cases	1.27	3.04	No
	The patient has severe disease	**3.18**	**2.22**	**For**
Specific non-biologics				

132. For most patients, methotrexate (MTX) should be the first non-biologic used if it is not contraindicated	**3.27**	**2.27**	**For**
133. For most patients, methotrexate (MTX) should be administered orally	**3.73**	**2.10**	**For**
134. Methotrexate injection may be preferable over oral administration to reduce nausea and GI adverse effects	**2.59**	**2.34**	**For**
135. Methotrexate injection may be preferable over oral administration to improve efficacy by avoiding the need for GI absorption	0.95	2.84	No
136. For most patients, azathioprine should be the first non-biologic used	-1.41	2.09	No
137. For most patients, azathioprine should be tried if MTX has failed or is not tolerated	1.91	2.56	No
138. For most patients, mycophenolate should be reserved for 3rd-line therapy	1.36	2.66	No
139. For most patients, leflunomide should be reserved for 3rd-line therapy	-0.41	3.17	No
140. Hydroxychloroquine may be useful in managing:			
Hypercalcemia	**2.55**	**1.68**	**For**
Skin disease	**3.41**	**1.33**	**For**
Bone/joint disease	2.05	1.70	No
Mild lung disease manifestations	-0.14	2.68	No
To manage fatigue	0.18	2.87	No
As a steroid-sparing therapy	1.05	2.15	No
141. There is little role for chlorambucil in managing pulmonary sarcoidosis	2.55	3.02	No
Combination therapies			
142. Combination therapy with two different non-biologics should be considered:			
To provide synergistic effects	1.64	2.61	No
To improve response when a single drug gives only a partial response	1.95	2.63	No
To avoid use of TNF antagonists	-0.27	3.03	No
If cardiac sarcoidosis is present	0.36	2.57	No
If neurosarcoidosis is present	0.18	2.59	No
If ocular sarcoidosis is present	0.18	2.63	No
If skin involvement is present	-0.14	2.49	No
Adjusting non-biologics			
143. If toxicity to a non-biologic develops, the option to consider should usually be:			

	Switching to a different non-biologic	**3.55**	**1.18**	**For**
	Switching to a biologic	2.18	2.15	No
	Reducing the dose	**2.73**	**2.07**	**For**
	Discontinuing the non-biologic	**3.00**	**1.38**	**For**
144.	If a patient is unable to tolerate a non-biologic, the option to consider should be:			
	Switching to a different non-biologic	**3.82**	**1.01**	**For**
	Switching to a biologic	**2.64**	**1.89**	**For**
	Reducing the dose	2.09	2.45	No
	Discontinuing the non-biologic	**3.55**	**1.41**	**For**
145.	If a patient is unable to manage the cost of a non-biologic, the option to consider should be:			
	Switching to a less expensive non-biologic	**3.32**	**1.67**	**For**
	Switching to a biologic	-0.32	2.57	No
	Reducing the dose	-0.27	2.60	No
	Discontinuing the non-biologic	1.50	2.39	No
146.	If a treatment failure occurs with a non-biologic, the option to consider should be:			
	Add a different non-biologic	0.36	2.57	No
	Switching to different non-biologic	2.41	2.44	No
	Switching to a biologic	**3.45**	**1.71**	**For**
	Reducing the dose	-2.91	2.49	Against
	Discontinuing the non-biologic	**2.64**	**2.28**	**For**
147.	If the patient prefers a different non-biologic, the option to consider should be:			
	Add a different non-biologic	-0.27	2.57	No
	Switching to different non-biologic	**3.18**	**1.40**	**For**
148.	Hydroxychloroquine has a role in combination with other non-biologics for some patients	2.09	1.87	No
149.	Folic acid has a role in combination with other non-biologics for some patients	2.00	2.51	No
150.	Discontinuation of a non-biologic therapy should be considered for:			
	Toxicity	**4.50**	**0.96**	**For**
	Intolerability	**4.55**	**0.67**	**For**
	Failure to stabilize disease	**3.95**	**1.70**	**For**

Long-term disease stabilization	**2.50**	**2.28**	**For**
Failure to withdraw /substantially decrease oral corticosteroids	1.77	2.29	No
Biologic Therapies			
151. Biologic therapies should be considered if:			
Steroids are toxic or not sufficiently effective	1.73	2.76	No
Non-biologics are toxic or not sufficiently effective	**3.73**	**1.24**	**For**
Steroids and non-biologics in combination are toxic or not sufficiently effective	**4.50**	**0.60**	**For**
Severe or progressive disease is present	**3.73**	**1.42**	**For**
152. For most patients, infliximab should be the first biologic used if it is not contraindicated	**3.73**	**1.32**	**For**
153. For most patients, a TNF inhibitor should be the first biologic used if it is not contraindicated	**4.50**	**0.67**	**For**
154. Repository corticotrophin injection (RCI) has a role for some patients	0.86	3.47	No
Infliximab dosing			
155. Most patients should receive a loading dose when infliximab is started	**2.64**	**2.61**	**For**
156. For most patients, the infliximab loading dose should be 3-6 mg/kg at weeks 0, 2, and 6	1.91	2.97	No
157. For most patients, the infliximab loading dose should be 5 mg/kg at weeks 0, 2, and 6	**2.86**	**2.32**	**For**
158. For most patients, the infliximab loading dose should be 3 mg/kg at weeks 1, 2, and 4	-0.09	2.83	No
159. For most patients, the infliximab loading dose should be 6 mg/kg on days 1 and 15	-0.77	2.51	No
160. For most patients, the infliximab maintenance dose should be 5 mg/kg every 4-8 weeks	1.91	2.84	No
161. For most patients, the infliximab maintenance dose should be 5 mg/kg every 4 weeks	2.23	2.43	No
162. For most patients, the infliximab maintenance dose should be 5 mg/kg every 6 weeks	0.59	2.89	No
163. For most patients, the infliximab maintenance dose should be 5 mg/kg every 8 weeks	-0.27	2.81	No
Adalimumab dosing			
164. Most patients should receive a loading dose when adalimumab is started	1.56	2.50	No
165. For most patients, the adalimumab loading dose should be 160 mg at week 0 and 80 mg at week 2	1.39	2.35	No
166. For most patients, the adalimumab loading dose should be 120 mg at week 0 and 80 mg at week 2	0.22	2.26	No
167. For most patients, the adalimumab loading dose should be 80 mg weekly for 2 weeks	-0.11	2.14	No
168. For most patients, the adalimumab maintenance dose should be 40-80 mg every other week	1.22	2.24	No
169. For most patients, the adalimumab maintenance dose should be 40 mg every other week	1.83	2.15	No
170. For most patients, the adalimumab maintenance dose should be 40 mg every week	2.00	2.22	No

Rituximab dosing			
171. Rituximab is appropriate as an alternative third-line therapy if anti-TNF therapy is contraindicated or ineffective	1.50	2.74	No
172. Most patients should receive a loading dose when rituximab is started	1.18	2.70	No
173. For most patients, the rituximab loading dose should 1000 mg on days 1 and 15	1.95	2.17	No
174. For most patients, the rituximab dose should be one course every 24 weeks. One course is two 1000-mg IV infusions separated by 2 weeks	0.91	2.56	No
175. For most patients, the rituximab maintenance dose should be 1000 mg every 3-5 weeks	-0.50	2.86	No
176. For most patients, the rituximab maintenance dose should be 500 mg	-0.41	2.50	No
Repository corticotrophin injection (RCI) dosing			
177. For most patients receiving RCI, the regimen should be 40-80 IU every 24-72 hours	0.53	2.97	No
178. For most patients receiving RCI, the regimen should be 40 IU twice weekly	1.05	2.97	No
179. For most patients receiving RCI, the regimen should be 80 IU every other week	-1.53	2.29	No
180. For most patients receiving RCI, the regimen should be 80 IU, then 40 IU every 72 hours	-0.21	2.53	No
181. For most patients receiving RCI, the regimen should be 40 IU twice weekly, 80 IU twice weekly, or 80 IU three times weekly depending on patient response	0.32	2.83	No
182. The RCI dose should be titrated to achieve a response	0.95	2.72	No
183. Most patients should receive a loading dose when RCI is started	-1.32	2.40	No
Combination Therapies			
184. For most patients, biologics should be used in combination with steroids	0.73	3.10	No
185. For most patients, biologics should be used in combination with non-biologics	**2.50**	**2.28**	**For**
186. For most patients, biologics should be used in combination with steroids and non-biologics	-0.23	2.76	No
187. When biologics are used with steroids, a low-dose steroid regimen is appropriate	**3.27**	**2.07**	**For**
188. For most patients, biologics should be used in combination with MTX	**2.64**	**1.99**	**For**
189. When biologics are used with MTX, a low-dose MTX regimen is appropriate	**3.55**	**1.60**	**For**
190. The combination of biologics and MTX is beneficial because it reduces the risk of autoantibody formation	**3.50**	**1.47**	**For**
191. For most patients, biologics should be used in combination with azathioprine	-0.77	2.22	No
Discontinuing Biologics			
192. In a patient who has initially responded to biologics, discontinuation of the biologic is appropriate if:			

Toxicity has developed	**4.14**	**1.93**	**For**
The patient finds therapy is too costly	2.27	2.78	No
Therapy has failed to stabilize the disease	**3.73**	**1.91**	**For**
The disease has been stable for at least 6-12 months	0.14	3.09	No
The disease has been stable for at least 1-2 years	2.32	1.91	No
The disease has been stable for at least 2-3 years	**3.64**	**1.36**	**For**
193. When discontinuing biologics, the patient should be weaned by:			
Increasing the interval between biologic doses	2.41	2.52	No
Allowing rheumatology to manage discontinuation	-2.14	3.03	No
194. Antimicrobial prophylaxis for pneumocystis pneumonia (PCP) should be used:			
For all patients receiving biologics	-0.50	2.72	No
Patients at risk for infection	**2.73**	**2.37**	**For**
High-dose immunosuppression with multiple agents	**2.86**	**2.46**	**For**
Prolonged high-dose steroids	2.77	3.15	No
Never	-3.14	2.25	Against
195. The following risk factors should be considered in the decision to use antimicrobial prophylaxis for PCP:			
Other comorbidities such as diabetes and ESRD	1.95	2.52	No
Prior infection	**3.23**	**1.69**	**For**
Use of multiple agents	**2.77**	**1.80**	**For**
Low CD4 counts	2.14	2.29	No
High-dosage prednisone	2.45	3.33	No
196. Antimicrobial prophylaxis for tuberculosis (TB) should be used:			
For all patients receiving biologics	-2.95	2.10	Against
In patients with a positive PPD test	2.14	2.82	No
In patients with a positive interferon gamma test	**2.77**	**2.56**	**For**
In patients with a history of TB or latent TB	**2.59**	**2.54**	**For**
Prolonged high-dose steroids	-2.86	2.25	Against
Never	-3.73	1.78	Against

197. Screening for latent tuberculosis should use:			
PPD	1.55	2.69	No
IFN-gamma testing	**4.27**	**0.88**	**For**
Chest x-ray changes	1.50	3.35	No
198. Most patients should be vaccinated for pneumococcal disease and influenza	**4.05**	**2.26**	**For**
Additional therapies			
199. Pentoxifylline may be appropriate as a “last resort” therapy	-1.00	3.32	No
200. Thalidomide has a role for some patients	-0.15	3.36	No
201. There is little role for chloroquine in managing pulmonary sarcoidosis	1.67	2.82	No
202. There is little role for cyclosporine in managing pulmonary sarcoidosis	1.36	3.13	No
203. There is little role for colchicine in managing pulmonary sarcoidosis	**3.05**	**2.08**	**For**
204. There is little role for CLEAR (concomitant levofloxacin, ethambutol, azithromycin, rifampin) in managing pulmonary sarcoidosis	1.62	3.01	No
205. Lung transplantation should be considered:			
Severe disease not responsive to therapy	**4.18**	**1.74**	**For**
Pulmonary hypertension	**3.59**	**2.44**	**For**
Low and worsening PFTs	**3.77**	**2.00**	**For**
206. Comment (if needed) • Q83. Very difficult to know how to answer this question. Cardiac MRI, PET, echo, Holter, SR, vitamin D may be of importance but is not performed in every patient at presentation in our clinic, so it is difficult to know how to answer. I don´t know what is included in CMP. • Modifications of ratings result from: - rare true errors (for ex: 65 for +5) - limiting my trend to strong responses (down-grading from 5 to 5 for ex) - probably educational response (I had first interpreted some questions as exclusive from the following • Some questions were difficult to answer due to range of possibilities (e.g., reduce prednisone dose in diabetic/obese- depends on severity of comorbidities) (when to use combination steroids/non-biologics/biologics really depends on individual patient, etc)			

DLCO, diffusing capacity of the lungs for carbon monoxide; FVC, forced vital capacity; HRCT, high-resolution computed tomography; QOL, quality of life.