

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

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American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease

Project Plan – August 2022

1 **ORGANIZATIONAL LEADERSHIP AND SUPPORT**

2
3 This project is led and funded by the American College of Rheumatology (ACR).
4

5 **BACKGROUND**

6 Interstitial lung disease (ILD), a heterogeneous group of disorders characterized by inflammation
7 and fibrosis of the lung parenchyma, is a significant cause of morbidity and mortality in people
8 with systemic autoimmune rheumatic diseases (ARDs). Although all people with ARDs are at risk
9 for developing ILD, those with systemic sclerosis (SSc), rheumatoid arthritis (RA), mixed
10 connective tissue disease (MCTD), polymyositis/dermatomyositis (PM/DM), and Sjogren’s
11 Syndrome (SS) are at the greatest risk (1,2) For example, ILD affects approximately 40-60% of
12 adults with SSc and is the leading cause of death and hospitalization in this population (3-6).
13

14 Despite the life-threatening nature of ARD-ILD and the emerging potential of new therapies to
15 arrest disease progression, there are no clinical practice guidelines for ILD screening or
16 treatment in the ARDs. It is hypothesized that screening for ARD-ILD could lead to early
17 interventions to prevent or slow progression of this often lethal disease. Hence, there is an
18 urgent need to develop ARD-ILD screening, monitoring, and treatment guidelines.
19
20

21 **OBJECTIVES**

22
23 The objective of this project is to develop a clinical practice guideline that includes evidence-
24 based consensus recommendations for clinicians who care for people with systemic
25 autoimmune rheumatic disease who are at risk for or have been diagnosed with interstitial lung
26 disease (ILD).
27

28 Specifically, we aim to:

- 29 1. Develop recommendations regarding optimal screening tests to screen for ILD in people
30 with specific ARDs.
31 2. Develop recommendations for the monitoring of ARD-ILD (monitoring for both the
32 development and progression of ARD-ILD).
33 3. Develop treatment recommendations for ARD-ILD.
34

35 **THEMES OF PICO QUESTIONS**

36 The PICO (Population/patients, Intervention, Comparator, and Outcomes; *see Appendix A*) questions
37 developed for this guideline fall into 5 major categories:

American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease

Project Plan – August 2022

- 38 1. Screening for ILD in people with rheumatic disease at increased risk of developing ILD
39 2. Monitoring ILD progression and treatment complications
40 3. Treatment of ILD: first therapy
41 4. Treatment of ILD after ILD progression on first ILD therapy
42 5. Treatment of rapidly progressive ILD

43

44 **METHODS**

45

46 *Identification of Studies*

47 Literature search strategies, based on PICO questions, were drafted by a research librarian with input
48 from the Core Team. Searches were performed in OVID Medline (1946 +), Embase (1974 +), and
49 PubMed (mid-1960s +).

50

51 The search strategies were developed using the controlled vocabulary or thesauri language for each
52 database: Medical Subject Headings (MeSH) for OVID Medline and PubMed; and Emtree terms for
53 Embase. Text words were also used in OVID Medline, PubMed, and Embase.

54

55 *Search Limits*

56 Only English language articles will be retrieved.

57

58 *Literature Search Update*

59 Literature searches will be updated just before the voting panel meeting to ensure completeness.

60

61 *Inclusion/Exclusion Criteria*

62 *Appendix A* includes the project's PICO questions, which outline the defined patient population,
63 interventions, comparators, and outcomes. *Appendix B* includes the list of inclusion/exclusion criteria.

64

65 *Management of Studies and Data*

66 References and abstracts will be imported into bibliographic management software (EndNote) (7),
67 duplicates removed, and exported to Distiller SR, a web-based systematic review manager (8). Screening
68 and data abstraction forms will be created in Distiller SR. Search results will be divided among reviewers,
69 and two reviewers will screen each title/abstract, with disagreements at the title/abstract screening
70 stage defaulting to inclusion for full manuscript review. Following the same dual review process,
71 disagreements at the full manuscript screening stage will be discussed and adjudicated by the literature
72 review leadership, if necessary.

73

74 *Phases*

- 75 1. A search for randomized controlled trials and observational studies will be performed to
76 determine existing studies covering outcomes of interest.

American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease

Project Plan – August 2022

- 77 2. Additionally, recently published systematic reviews covering outcomes of interest will also be
78 sought and used for reference cross-checking.
79 3. Chosen studies will be quality-assessed using the Instrument to assess the Credibility of Effect
80 Modification Analyses.
81 4. Subsequently, identified studies will be assessed using the RevMan (10) and GRADE Pro tools
82 (11).

83

84 *GRADE Methodology*

85

86 GRADE methodology will be used in this project to grade available evidence and facilitate development
87 of recommendations. The certainty in the evidence (also known as ‘quality’ of evidence) will be graded
88 as high, moderate, low or very low. The recommendations will have a strength, strong or conditional,
89 and a direction, as in favor of or against the intervention. The strength of recommendations will not
90 depend solely on the certainty in the evidence, but also on patient preferences and values, and the
91 weight between benefits and harms. A series of articles that describe the GRADE methodology can be
92 found on the GRADE working group’s website: www.gradeworkinggroup.org.

93

94 *Data Analysis and Synthesis*

95

96 The literature review team will analyze and synthesize data from included studies that address the PICO
97 questions. An evidence profile, including a GRADE Summary of Findings table, will be prepared for each
98 PICO question using Review Manager (RevMan) (10) and GRADEprofiler (GRADEpro) software (11). The
99 Summary of Findings table contains the benefits and harms for each outcome across studies, the
100 assumed and corresponding risk for comparators and interventions (95% CI), the absolute risk and
101 relative effect (95% CI), the number of participants/number of studies, and the certainty in the evidence
102 for each critical and important outcome (i.e., high, moderate, low or very low).

103

104 The evidence profile documents the overall certainty in the evidence for each critical and important
105 outcome across studies and summarizes the rationale of the GRADE criteria for downgrading (risk of
106 bias, inconsistency, indirectness, imprecision, and publication bias), or upgrading the certainty in a body
107 of evidence (large magnitude of effect, dose-response gradient, and all plausible confounding that
108 would reduce a demonstrated effect).

109

110 *Development of Recommendation Statements*

111

112 PICO questions will be revised into drafted recommendation statements. Using the GRADE Evidence
113 Profiles and Summaries of Findings tables, the voting panel, consisting of 11 rheumatologists, 5
114 pulmonologists, 1 radiologist, and at least 2 patients with rheumatic disease-associated ILD (specifically,
115 ILD associated with either systemic sclerosis, rheumatoid arthritis, mixed connective tissue disease,
116 dermatomyositis, polymyositis, or Sjogren’s syndrome), will consider the drafted recommendation
117 statements in two stages. The first assessment will be done individually, and the results will be

American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease

Project Plan – August 2022

118 anonymous; this vote will only be used to determine where consensus might or might not already exist
119 and develop an agenda for a virtual voting panel meeting. At the virtual voting panel meeting, chaired by
120 the principal investigators, the panelists will discuss the evidence in the context of their clinical
121 experience and expertise to arrive at consensus on the final recommendations. The voting panel meeting
122 discussions will be supported by the GRADE expert as well as the literature review leader, who will attend
123 the meeting to provide details about the evidence, as requested. Voting panel discussions and decisions
124 will also be informed by a separately convened patient panel, which will meet in the days before the
125 voting panel meeting, to provide unique patient perspectives on the drafted recommendations based on
126 their experiences and the available literature.

127

128 **PLANNED APPENDICES (AT MINIMUM)**

129

130 A. Final literature search strategies

131 B. Inclusion/exclusion criteria

132 C. GRADE evidence profiles and summary of findings tables for each PICO question

133

134 **AUTHORSHIP**

135

136 Authorship of the guideline will include co-principal investigators Sindhu R. Johnson, MD, PhD, and Elana
137 J. Bernstein, MD, MSc; co-literature review leaders Ilya Ivlev, MD, PhD, MBI and Stacey Uhl; MS content
138 experts Marcy B. Bolster, MD, Jonathan H. Chung, MD, Sonye Danoff, MD, PhD, Michael George, MD,
139 MSCE, and Dinesh Khanna, MD; and Gordon Guyatt, MD, GRADE expert, and Reza Mirza, MD,
140 GRADE/Methodological Contributor. Members of the voting panel and literature review team will also
141 be authors. The PIs will determine final authorship, dependent on the efforts made by individuals
142 throughout the guideline development process, using international authorship standards as guidance.

143

144 **DISCLOSURES/CONFLICTS OF INTEREST**

145

146 The ACR's disclosure and COI policies for guideline development will be followed for this project. These
147 can be found in the ACR Guideline Manual on [this page of the ACR web site](#), under Policies &
148 Procedures. *See Appendix D for participant disclosures.*

149

150

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152

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American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease

Project Plan – August 2022

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**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

177 **ARDs included in this project**

- 178 **1. Systemic sclerosis**
179 **2. Rheumatoid arthritis**
180 **3. Inflammatory myopathy**
181 **4. Sjogren’s Syndrome**
182 **5. Mixed connective tissue disease**

183

184

185 **SUMMARY OF PICO QUESTIONS**

186

187 **Screening PICO questions:**

188 In people with ARD at increased risk of developing ILD, what is the impact of each of the following tests on
189 diagnostic accuracy, disease-related outcomes, and diagnostic testing-related adverse events?

- 190 • PFTs vs. history/physical
191 • High resolution CT thorax vs. history/physical
192 • 6-minute walk test distance vs. history/physical
193 • Chest radiograph vs. history/physical
194 • Ambulatory desaturation vs. history/physical
195 • Chest radiograph vs. high resolution CT thorax
196 • PFTs vs. ambulatory desaturation
197 • High resolution CT thorax vs. PFTs
198 • High resolution CT thorax and PFTs vs. PFTs alone
199 • Bronchoscopy vs. no bronchoscopy
200 • Surgical lung biopsy vs. no surgical lung biopsy

201

202 **Monitoring PICO questions:**

203 In people with ARD who also have ILD, what is the impact of each of the following tests on
204 responsiveness/sensitivity to change of the test, disease-related outcomes, treatment-related serious adverse
205 events and testing-related adverse events?

- 206 • PFTs vs. history/physical
207 • High resolution CT thorax vs. history/physical
208 • 6-minute walk test distance vs. history/physical
209 • Chest radiograph vs. history/physical
210 • Ambulatory desaturation vs. history/physical
211 • Chest radiograph vs. high resolution CT thorax
212 • Bronchoscopy vs. no bronchoscopy

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

- 213 • High resolution CT thorax vs. bronchoscopy
214 • PFTs vs. 6-minute walk test distance
215 • PFTs and 6-minute walk test distance vs. PFTs alone
216 • PFTs vs. ambulatory desaturation
217 • PFTs and high resolution CT thorax vs. PFTs alone
218 • 6-minute walk test distance vs. ambulatory desaturation

219

220 **Medical management – 1st ILD therapy PICO questions:**

221 In people with ARD who also have ILD, what is the impact of each of the following therapies as first line ILD
222 treatment on disease-related outcomes and treatment-related adverse events?

- 223 • Mycophenolate vs. no mycophenolate
224 • Cyclophosphamide vs. no cyclophosphamide (I/V or oral)
225 • Leflunomide vs. no leflunomide
226 • Methotrexate vs. no methotrexate
227 • Azathioprine vs. no azathioprine
228 • Calcineurin inhibitors vs. no calcineurin inhibitors
229 • Anti-TNF therapy vs. no anti-TNF therapy
230 • Abatacept vs. no abatacept
231 • Anti-CD20 antibody vs. no anti-CD20 antibody
232 • IL-6 receptor antagonists vs. no IL-6 receptor antagonists
233 • JAK inhibitors vs. no JAK inhibitors
234 • Daily oral prednisone vs. no daily oral prednisone
235 • IV pulse glucocorticoids vs. no IV pulse glucocorticoids
236 • Nintedanib vs. no nintedanib
237 • Pirfenidone vs. no pirfenidone
238 • IVIG vs. no IVIG
239 • Plasma exchange vs. no plasma exchange
240 • Adding nintedanib to mycophenolate vs. not adding nintedanib to mycophenolate
241 • Adding pirfenidone to mycophenolate vs. not adding pirfenidone to mycophenolate
242 • Upfront combination of nintedanib with mycophenolate vs. mycophenolate alone
243 • Upfront combination of pirfenidone with mycophenolate vs. mycophenolate alone
244 • Methotrexate vs. mycophenolate
245 • Leflunomide vs. mycophenolate
246 • Azathioprine vs. mycophenolate
247 • I/V or oral cyclophosphamide vs. mycophenolate
248 • Calcineurin inhibitors vs. mycophenolate

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

- 249 • TNF inhibitors vs. mycophenolate
- 250 • IL-6 receptor antagonists vs. mycophenolate
- 251 • Anti-CD20 antibody vs. mycophenolate
- 252 • Abatacept vs. mycophenolate
- 253 • JAK inhibitors vs. mycophenolate
- 254 • Nintedanib vs. mycophenolate
- 255 • Pirfenidone vs. mycophenolate
- 256 • IVIG vs. mycophenolate
- 257 • Oral prednisone vs. mycophenolate
- 258 • Intravenous methylprednisolone vs. mycophenolate
- 259 • Plasma exchange vs. mycophenolate
- 260 • Methotrexate vs. anti-CD20 antibody
- 261 • Leflunomide vs. anti-CD20 antibody
- 262 • Azathioprine vs. anti-CD20 antibody
- 263 • I/V or oral cyclophosphamide vs. anti-CD20 antibody
- 264 • Calcineurin inhibitors vs. anti-CD20 antibody
- 265 • TNF inhibitors vs. anti-CD20 antibody
- 266 • IL-6 receptor antagonists vs. anti-CD20 antibody
- 267 • Abatacept vs. anti-CD20 antibody
- 268 • JAK inhibitors vs. anti-CD20 antibody
- 269 • Nintedanib vs. anti-CD20 antibody
- 270 • Pirfenidone vs. anti-CD20 antibody
- 271 • IVIG vs. anti-CD20 antibody
- 272 • Oral prednisone vs. anti-CD20 antibody
- 273 • Intravenous methylprednisolone vs. anti-CD20 antibody
- 274 • Plasma exchange vs. anti-CD20 antibody
- 275 • Methotrexate vs. azathioprine
- 276 • Leflunomide vs. azathioprine
- 277 • I/V or oral cyclophosphamide vs. azathioprine
- 278 • Calcineurin inhibitors vs. azathioprine
- 279 • TNF inhibitors vs. azathioprine
- 280 • IL-6 receptor antagonists vs. azathioprine
- 281 • Abatacept vs. azathioprine
- 282 • JAK inhibitors vs. azathioprine
- 283 • Nintedanib vs. azathioprine

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

- 284 • Pirfenidone vs. azathioprine
- 285 • IVIG vs. azathioprine
- 286 • Oral prednisone vs. azathioprine
- 287 • Intravenous methylprednisolone vs. azathioprine
- 288 • Plasma exchange vs. azathioprine
- 289 • Methotrexate vs. I/V or oral cyclophosphamide
- 290 • Leflunomide vs. I/V or oral cyclophosphamide
- 291 • Calcineurin inhibitors vs. I/V or oral cyclophosphamide
- 292 • TNF inhibitors vs. I/V or oral cyclophosphamide
- 293 • IL-6 receptor antagonists vs. I/V or oral cyclophosphamide
- 294 • Abatacept vs. I/V or oral cyclophosphamide
- 295 • JAK inhibitors vs. I/V or oral cyclophosphamide
- 296 • Nintedanib vs. I/V or oral cyclophosphamide
- 297 • Pirfenidone vs. I/V or oral cyclophosphamide
- 298 • IVIG vs. I/V or oral cyclophosphamide
- 299 • Oral prednisone vs. I/V or oral cyclophosphamide
- 300 • Intravenous methylprednisolone vs. I/V or oral cyclophosphamide
- 301 • Plasma exchange vs. I/V or oral cyclophosphamide
- 302 • Nintedanib vs. IL-6 receptor antagonists
- 303 • Referral for stem cell transplant vs. optimal medical management
- 304 • Referral for lung transplant vs. optimal medical management

305

306 Medical management – ILD progression on any 1st ILD therapy PICO questions

307 In ARD patients with ILD progression after 1st ILD therapy, what is the impact of adding each of the following
308 therapies on disease-related outcomes and treatment-related adverse events?

- 309 • Combination of nintedanib and mycophenolate vs. mycophenolate alone
- 310 • Combination of pirfenidone and mycophenolate vs. mycophenolate alone
- 311 • Methotrexate vs. mycophenolate
- 312 • Leflunomide vs. mycophenolate
- 313 • Azathioprine vs. mycophenolate
- 314 • I/V or oral cyclophosphamide vs. mycophenolate
- 315 • Calcineurin inhibitors vs. mycophenolate
- 316 • TNF inhibitors vs. mycophenolate
- 317 • IL-6 receptor antagonists vs. mycophenolate
- 318 • Anti-CD20 antibody vs. mycophenolate
- 319 • Abatacept vs. mycophenolate

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

- 320 • JAK inhibitors vs. mycophenolate
- 321 • Nintedinib vs. mycophenolate
- 322 • Pirfenidone vs. mycophenolate
- 323 • IVIG vs. mycophenolate
- 324 • Oral prednisone vs. mycophenolate
- 325 • Intravenous methylprednisolone vs. mycophenolate
- 326 • Plasma exchange vs. mycophenolate
- 327 • Methotrexate vs. anti-CD20 antibody
- 328 • Leflunomide vs. anti-CD20 antibody
- 329 • Azathioprine vs. anti-CD20 antibody
- 330 • I/V or oral cyclophosphamide vs. anti-CD20 antibody
- 331 • Calcineurin inhibitors vs. anti-CD20 antibody
- 332 • TNF inhibitors vs. anti-CD20 antibody
- 333 • IL-6 receptor antagonists vs. anti-CD20 antibody
- 334 • Abatacept vs. anti-CD20 antibody
- 335 • JAK inhibitors vs. anti-CD20 antibody
- 336 • Nintedinib vs. anti-CD20 antibody
- 337 • Pirfenidone vs. anti-CD20 antibody
- 338 • IVIG vs. anti-CD20 antibody
- 339 • Oral prednisone vs. anti-CD20 antibody
- 340 • Intravenous methylprednisolone vs. anti-CD20 antibody
- 341 • Plasma exchange vs. anti-CD20 antibody
- 342 • Methotrexate vs. azathioprine
- 343 • Leflunomide vs. azathioprine
- 344 • I/V or oral cyclophosphamide vs. azathioprine
- 345 • Calcineurin inhibitors vs. azathioprine
- 346 • TNF inhibitors vs. azathioprine
- 347 • IL-6 receptor antagonists vs. azathioprine
- 348 • Abatacept vs. azathioprine
- 349 • JAK inhibitors vs. azathioprine
- 350 • Nintedinib vs. azathioprine
- 351 • Pirfenidone vs. azathioprine
- 352 • IVIG vs. azathioprine
- 353 • Oral prednisone vs. azathioprine
- 354 • Intravenous methylprednisolone vs. azathioprine

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

- 355 • Plasma exchange vs. azathioprine
- 356 • Methotrexate vs. I/V or oral cyclophosphamide
- 357 • Leflunomide vs. I/V or oral cyclophosphamide
- 358 • Calcineurin inhibitors vs. I/V or oral cyclophosphamide
- 359 • TNF inhibitors vs. I/V or oral cyclophosphamide
- 360 • IL-6 receptor antagonists vs. I/V or oral cyclophosphamide
- 361 • Abatacept vs. I/V or oral cyclophosphamide
- 362 • JAK inhibitors vs. I/V or oral cyclophosphamide
- 363 • Nintedanib vs. I/V or oral cyclophosphamide
- 364 • Pirfenidone vs. I/V or oral cyclophosphamide
- 365 • IVIG vs. I/V or oral cyclophosphamide
- 366 • Oral prednisone vs. I/V or oral cyclophosphamide
- 367 • Intravenous methylprednisolone vs. I/V or oral cyclophosphamide
- 368 • Plasma exchange vs. I/V or oral cyclophosphamide
- 369 • Referral for stem cell transplant vs. optimal medical management
- 370 • Referral for lung transplant vs. optimal medical management

371
372 **Medical management – rapidly progressive ILD PICO questions**

- 373 In ARD patients with rapidly progressive ILD, what is the impact of each of the following therapies as first line
374 rapidly progressive ILD treatment on disease-related outcomes and treatment-related adverse events?
- 375 • Daily oral prednisone vs. no daily oral prednisone
 - 376 • Pulse intravenous glucocorticoids vs. no pulse intravenous glucocorticoids
 - 377 • Nintedanib vs. no nintedanib
 - 378 • Pirfenidone vs. no pirfenidone
 - 379 • Adding nintedanib to mycophenolate vs. not adding nintedanib to mycophenolate
 - 380 • Adding pirfenidone to mycophenolate vs. not adding pirfenidone to mycophenolate
 - 381 • Upfront combination of nintedanib with mycophenolate vs. mycophenolate alone
 - 382 • Upfront combination of pirfenidone with mycophenolate vs. mycophenolate alone
 - 383 • Methotrexate vs. mycophenolate
 - 384 • Leflunomide vs. mycophenolate
 - 385 • Azathioprine vs. mycophenolate
 - 386 • I/V or oral cyclophosphamide vs. mycophenolate
 - 387 • Calcineurin inhibitors vs. mycophenolate
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 - 390 • Anti-CD20 antibody vs. mycophenolate

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

- 391 • Abatacept vs. mycophenolate
- 392 • JAK inhibitors vs. mycophenolate
- 393 • Nintedinib vs. mycophenolate
- 394 • Pirfenidone vs. mycophenolate
- 395 • IVIG vs. mycophenolate
- 396 • Oral prednisone vs. mycophenolate
- 397 • Intravenous methylprednisolone vs. mycophenolate
- 398 • Plasma exchange vs. mycophenolate
- 399 • Methotrexate vs. anti-CD20 antibody
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- 414 • Methotrexate vs. azathioprine
- 415 • Leflunomide vs. azathioprine
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- 420 • Abatacept vs. azathioprine
- 421 • JAK inhibitors vs. azathioprine
- 422 • Nintedinib vs. azathioprine
- 423 • Pirfenidone vs. azathioprine
- 424 • IVIG vs. azathioprine
- 425 • Oral prednisone vs. azathioprine

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

- 426 • Intravenous methylprednisolone vs. azathioprine
- 427 • Plasma exchange vs. azathioprine
- 428 • Methotrexate vs. cyclophosphamide
- 429 • Leflunomide vs. cyclophosphamide
- 430 • Calcineurin inhibitors vs. I/V or oral cyclophosphamide
- 431 • TNF inhibitors vs. I/V or oral cyclophosphamide
- 432 • of IL-6 receptor antagonists vs. I/V or oral cyclophosphamide
- 433 • Abatacept vs. I/V or oral cyclophosphamide
- 434 • JAK inhibitors vs. I/V or oral cyclophosphamide
- 435 • Nintedanib vs. I/V or oral cyclophosphamide
- 436 • Pirfenidone vs. I/V or oral cyclophosphamide
- 437 • IVIG vs. I/V or oral cyclophosphamide
- 438 • Oral prednisone vs. I/V or oral cyclophosphamide
- 439 • Intravenous methylprednisolone vs. I/V or oral cyclophosphamide
- 440 • Plasma exchange vs. I/V or oral cyclophosphamide
- 441 • Dual combination therapy* vs. monotherapy†
- 442 • Triple combination therapy‡ vs. monotherapy†
- 443 • Triple combination therapy‡ vs. dual combination therapy*
- 444 • IVIG and/or plasma exchange in addition to monotherapy†, dual combination therapy*, or triple
- 445 combination therapy‡ vs. monotherapy†, dual combination therapy*, or triple combination therapy‡
- 446 alone
- 447 • Antifibrotic (e.g., nintedanib or pirfenidone) in addition to monotherapy†, dual combination therapy*,
- 448 or triple combination therapy‡ vs. monotherapy†, dual combination therapy*, or triple combination
- 449 therapy‡ alone
- 450 • Referral for stem cell transplant vs. optimal medical management
- 451 • Referral for lung transplant vs. optimal medical management
- 452
- 453
- 454

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

455 **APPENDIX A – PICO Questions**

456

457 **Screening for ILD in people with rheumatic disease at increased risk of developing ILD**

458

459 1. In people with rheumatic disease at increased risk of developing ILD, what is the impact of pulmonary
460 function tests (PFTs) compared to history/physical alone (e.g., shortness of breath (dyspnea), functional
461 class and physical examination: crackles on auscultation) on diagnostic accuracy, disease-related
462 outcomes, and diagnostic testing-related adverse events?
463

464

465 2. In people with rheumatic disease at increased risk of developing ILD, what is the impact of high
466 resolution CT thorax compared to history/physical alone (e.g., shortness of breath (dyspnea), functional
467 class and physical examination: crackles on auscultation) on diagnostic accuracy, disease-related
468 outcomes, and diagnostic testing-related adverse events?
469

470

471 3. In people with rheumatic disease at increased risk of developing ILD, what is the impact of 6-minute
472 walk test distance compared to history/physical alone (e.g., shortness of breath (dyspnea), functional
473 class and physical examination: crackles on auscultation) on diagnostic accuracy, disease-related
474 outcomes, and diagnostic testing-related adverse events?
475

476

477 4. In people with rheumatic disease at increased risk of developing ILD, what is the impact chest
478 radiograph compared to history/physical alone (e.g., shortness of breath (dyspnea), functional class and
479 physical examination: crackles on auscultation) on diagnostic accuracy, disease-related outcomes,
480 and diagnostic testing-related adverse events?
481

482

483 5. In people with rheumatic disease at increased risk of developing ILD, what is the impact of ambulatory
484 desaturation compared to history/physical alone (e.g., shortness of breath (dyspnea), functional class
485 and physical examination: crackles on auscultation) on diagnostic accuracy, disease-related outcomes,
486 and diagnostic testing-related adverse events?
487

488

489 6. In people with rheumatic disease at increased risk of developing ILD, what is the impact of chest
490 radiograph compared to high resolution CT thorax on diagnostic accuracy, disease-related outcomes,
491 and diagnostic testing-related adverse events?
492

493

494 7. In people with rheumatic disease at increased risk of developing ILD, what is the impact of pulmonary
495 function tests (PFTs) compared to ambulatory desaturation on diagnostic accuracy, disease-related
496 outcomes, and diagnostic testing-related adverse events?
497

498

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

- 492 8. In people with rheumatic disease at increased risk of developing ILD, what is the impact of high
493 resolution CT thorax compared to PFTs on diagnostic accuracy, disease-related outcomes,
494 and diagnostic testing-related adverse events?
495
- 496 9. In people with rheumatic disease at increased risk of developing ILD, what is the impact of high
497 resolution CT thorax and PFTs compared to PFTs alone on diagnostic accuracy, disease-related
498 outcomes, and diagnostic testing-related adverse events?
499
- 500 10. In people with rheumatic disease at increased risk of developing ILD, what is the impact of
501 bronchoscopy (may include broncho-alveolar lavage, transbronchial biopsy, cryobiopsy) compared to no
502 bronchoscopy (may include broncho-alveolar lavage, transbronchial biopsy, cryobiopsy) on diagnostic
503 accuracy, disease-related outcomes, and diagnostic testing-related adverse events?
504
- 505 11. In people with rheumatic disease at increased risk of developing ILD, what is the impact of surgical lung
506 biopsy compared to no surgical lung biopsy on diagnostic accuracy, disease-related outcomes,
507 and diagnostic testing-related adverse events?
508

509 **Monitoring disease progression and treatment complications**

- 510 12. In people with rheumatic disease with ILD, what is the impact of pulmonary function tests (PFTs)
511 compared to history/physical alone (e.g., shortness of breath (dyspnea), functional class and physical
512 examination: crackles on auscultation) on responsiveness/sensitivity to change of the test, disease-
513 related outcomes, treatment-related serious adverse events and testing-related adverse events?
514
- 515 13. In people with rheumatic disease with ILD, what is the impact of high resolution CT thorax compared to
516 history/physical alone (e.g., shortness of breath (dyspnea), functional class and physical examination:
517 crackles on auscultation) on responsiveness/sensitivity to change of the test, disease-related
518 outcomes, treatment-related serious adverse events and testing-related adverse events?
519
- 520 14. In people with rheumatic disease with ILD, what is the impact of 6-minute walk test distance compared
521 to history/physical alone (e.g., shortness of breath (dyspnea), functional class and physical examination:
522 crackles on auscultation) on responsiveness/sensitivity to change of the test, disease-related
523 outcomes, treatment-related serious adverse events and testing-related adverse events?
524
- 525 15. In people with rheumatic disease with ILD, what is the impact of chest radiograph compared to
526 history/physical alone (e.g., shortness of breath (dyspnea), functional class and physical examination:

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

- 527 crackles on auscultation) on responsiveness/sensitivity to change of the test, disease-related
528 outcomes, treatment-related serious adverse events and testing-related adverse events?
529
- 530 16. In people with rheumatic disease with ILD, what is the impact ambulatory desaturation compared to
531 history/physical alone (e.g., shortness of breath (dyspnea), functional class and physician examination:
532 crackles on auscultation) on responsiveness/sensitivity to change of the test, disease-related
533 outcomes, treatment-related serious adverse events and testing-related adverse events?
534
- 535 17. In people with rheumatic disease with ILD, what is the impact of chest radiograph compared to high
536 resolution CT thorax on diagnostic accuracy, disease-related outcomes, and diagnostic testing-related
537 adverse events?
538
- 539 18. In people with rheumatic disease with ILD, what is the impact of bronchoscopy (may include broncho-
540 alveolar lavage, transbronchial biopsy, cryobiopsy) compared to no bronchoscopy (may include
541 broncho-alveolar lavage, transbronchial biopsy, cryobiopsy) on responsiveness/ sensitivity to change of
542 the test, disease-related outcomes, treatment-related serious adverse events and testing-related
543 adverse events?
544
- 545 19. In people with rheumatic disease with ILD, what is the impact of high resolution CT thorax compared to
546 bronchoscopy (may include broncho-alveolar lavage, transbronchial biopsy, cryobiopsy) on
547 responsiveness/ sensitivity to change of the test, disease-related outcomes, treatment-related serious
548 adverse events and testing-related adverse events?
549
- 550 20. In people with rheumatic disease with ILD, what is the impact of PFTs compared to 6-minute walk test
551 distance on responsiveness/ sensitivity to change of the test, disease-related outcomes, treatment-
552 related serious adverse events and testing-related adverse events?
553
- 554 21. In people with rheumatic disease with ILD, what is the impact of PFTs and 6-minute walk test distance
555 compared to PFTs alone on responsiveness/ sensitivity to change of the test, disease-related
556 outcomes, treatment-related serious adverse events and testing-related adverse events?
557
- 558 22. In people with rheumatic disease with ILD, what is the impact of PFTs compared to ambulatory
559 desaturation on responsiveness/ sensitivity to change of the test, disease-related outcomes, treatment-
560 related serious adverse events and testing-related adverse events?
561

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

- 562 23. In people with rheumatic disease with ILD, what is the impact of PFTs and high resolution CT thorax
563 compared to PFTs alone on responsiveness/ sensitivity to change of the test, disease-related
564 outcomes, treatment-related serious adverse events and testing-related adverse events?
565
- 566 24. In people with rheumatic disease with ILD, what is the impact of 6-minute walk test distance compared
567 to ambulatory desaturation on responsiveness/ sensitivity to change of the test, disease-related
568 outcomes, treatment-related serious adverse events and testing-related adverse events?
569

570 **Medical Management**

571 **Rheumatic disease 1st ILD therapy**

- 572
- 573 25. In people with rheumatic disease with ILD, what is the impact of mycophenolate compared to no
574 mycophenolate as first line ILD treatment on disease-related outcomes and treatment-related adverse
575 events?
576
- 577 26. In people with rheumatic disease with ILD, what is the impact of cyclophosphamide compared to no
578 cyclophosphamide as first line ILD treatment on disease-related outcomes and treatment-related
579 adverse events?
580
- 581 27. In people with rheumatic disease with ILD, what is the impact of leflunomide compared to no
582 leflunomide as first line ILD treatment on disease-related outcomes and treatment-related adverse
583 events?
584
- 585 28. In people with rheumatic disease with ILD, what is the impact of methotrexate compared to no
586 methotrexate as first line ILD treatment on disease-related outcomes and treatment-related adverse
587 events?
588
- 589 29. In people with rheumatic disease with ILD, what is the impact of azathioprine compared to no
590 azathioprine as first line ILD treatment on disease-related outcomes and treatment-related adverse
591 events?
592
- 593 30. In people with rheumatic disease with ILD, what is the impact of calcineurin inhibitors compared to no
594 calcineurin inhibitors as first line ILD treatment on disease-related outcomes and treatment-related
595 adverse events?
596

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

- 597 31. In people with rheumatic disease with ILD, what is the impact of anti-TNF therapy compared to no anti-
598 TNF therapy as first line ILD treatment on disease-related outcomes and treatment-related adverse
599 events?
600
- 601 32. In people with rheumatic disease with ILD, what is the impact of abatacept compared to no abatacept
602 as first line ILD treatment on disease-related outcomes and treatment-related adverse events?
603
- 604 33. In people with rheumatic disease with ILD, what is the impact of anti-CD20 antibody (rituximab,
605 ocrelizumab, obinutuzumab, ofatumumab) compared to no anti-CD20 antibody (rituximab,
606 ocrelizumab, obinutuzumab, ofatumumab) as first line ILD treatment on disease-related outcomes and
607 treatment-related adverse events?
608
- 609 34. In people with rheumatic disease with ILD, what is the impact of IL-6 receptor antagonists (tocilizumab,
610 sarilumab) compared to no IL-6 receptor antagonists (tocilizumab, sarilumab) as first line ILD treatment
611 on disease-related outcomes and treatment-related adverse events?
612
- 613 35. In people with rheumatic disease with ILD, what is the impact of JAK inhibitors compared to no JAK
614 inhibitors as first line ILD treatment on disease-related outcomes and treatment-related adverse
615 events?
616
- 617 36. In people with rheumatic disease with ILD, what is the impact of daily oral prednisone compared to no
618 daily oral prednisone as first line ILD treatment on disease-related outcomes and treatment-related
619 adverse events?
620
- 621 37. In people with rheumatic disease with ILD, what is the impact of IV pulse glucocorticoids compared to
622 no IV pulse glucocorticoids first line ILD treatment on disease-related outcomes and treatment-related
623 adverse events?
624
- 625 38. In people with rheumatic disease with ILD, what is the impact of nintedanib compared to no nintedanib
626 as first line ILD treatment on disease-related outcomes and treatment-related adverse events?
627
- 628 39. In people with rheumatic disease with ILD, what is the impact of pirfenidone compared to no
629 pirfenidone as first line ILD treatment on disease-related outcomes and treatment-related adverse
630 events?
631

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

- 632 40. In people with rheumatic disease with ILD, what is the impact of IVIG compared to no IVIG as first line
633 ILD treatment on disease-related outcomes and treatment-related adverse events?
634
- 635 41. In people with rheumatic disease with ILD, what is the impact of plasma exchange compared to no
636 plasma exchange as first line ILD treatment on disease-related outcomes and treatment-related adverse
637 events?
638
- 639 42. In people with rheumatic disease with ILD without ILD progression, what is the impact of adding
640 nintedanib to mycophenolate compared to not adding nintedanib to mycophenolate on disease-related
641 outcomes and treatment-related adverse events?
642
- 643 43. In people with rheumatic disease with ILD without ILD progression, what is the impact of adding
644 pirfenidone to mycophenolate compared to not adding pirfenidone to mycophenolate on disease-
645 related outcomes and treatment-related adverse events?
646
- 647 44. In people with rheumatic disease with ILD, what is the impact of upfront combination of nintedanib
648 with mycophenolate compared to mycophenolate alone as first line ILD treatment on disease-related
649 outcomes and treatment-related adverse events?
650
- 651 45. In people with rheumatic disease with ILD, what is the impact of upfront combination of pirfenidone
652 with mycophenolate compared to mycophenolate alone as first line ILD treatment on disease-related
653 outcomes and treatment-related adverse events?
654
- 655 46. In people with rheumatic disease with ILD, what is the impact of methotrexate compared to
656 mycophenolate as first line ILD treatment on disease-related outcomes and treatment-related adverse
657 events?
658
- 659 47. In people with rheumatic disease with ILD, what is the impact of leflunomide compared to
660 mycophenolate as first line ILD treatment on disease-related outcomes and treatment-related adverse
661 events?
662
- 663 48. In people with rheumatic disease with ILD, what is the impact of azathioprine compared to
664 mycophenolate as first line ILD treatment on disease-related outcomes and treatment-related adverse
665 events?
666
- 667 49. In people with rheumatic disease with ILD, what is the impact of cyclophosphamide compared to
668 mycophenolate as first line ILD treatment on disease-related outcomes and treatment-related adverse

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

669 events?

670

671 50. In people with rheumatic disease with ILD, what is the impact of calcineurin inhibitors compared to
672 mycophenolate as first line ILD treatment on disease-related outcomes and treatment-related adverse
673 events?

674

675 51. In people with rheumatic disease with ILD, what is the impact of TNF inhibitors compared to
676 mycophenolate as first line ILD treatment on disease-related outcomes and treatment-related adverse
677 events?

678

679 52. In people with rheumatic disease with ILD, what is the impact of IL-6 receptor antagonists (tocilizumab,
680 sarilumab) compared to mycophenolate as first line ILD treatment on disease-related outcomes and
681 treatment-related adverse events?

682

683 53. In people with rheumatic disease with ILD, what is the impact of anti-CD20 antibody (rituximab,
684 ocrelizumab, obinutuzumab, ofatumumab) compared to mycophenolate as first line ILD treatment on
685 disease-related outcomes and treatment-related adverse events?

686

687 54. In people with rheumatic disease with ILD, what is the impact of abatacept compared to mycophenolate
688 as first line ILD treatment on disease-related outcomes and treatment-related adverse events?

689

690 55. In people with rheumatic disease with ILD, what is the impact of JAK inhibitors compared to
691 mycophenolate as first line ILD treatment on disease-related outcomes and treatment-related adverse
692 events?

693

694 56. In people with rheumatic disease with ILD, what is the impact of nintedinib compared to
695 mycophenolate as first line ILD treatment on disease-related outcomes and treatment-related adverse
696 events?

697

698 57. In people with rheumatic disease with ILD, what is the impact of pirfenidone compared to
699 mycophenolate as first line ILD treatment on disease-related outcomes and treatment-related adverse
700 events?

701

702 58. In people with rheumatic disease with ILD, what is the impact of IVIG compared to mycophenolate as
703 first line ILD treatment on disease-related outcomes and treatment-related adverse events?

704

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

- 705 59. In people with rheumatic disease with ILD, what is the impact of oral prednisone compared to
706 mycophenolate as first line ILD treatment on disease-related outcomes and treatment-related adverse
707 events?
708
- 709 60. In people with rheumatic disease with ILD, what is the impact of intravenous methylprednisolone
710 compared to mycophenolate as first line ILD treatment on disease-related outcomes and treatment-
711 related adverse events?
712
- 713 61. In people with rheumatic disease with ILD, what is the impact of plasma exchange compared to
714 mycophenolate as first line ILD treatment on disease-related outcomes and treatment-related adverse
715 events?
716
- 717 62. In people with rheumatic disease with ILD, what is the impact of methotrexate compared to anti-CD20
718 antibody (rituximab, ocrelizumab, obinutuzumab, ofatumumab) as first line ILD treatment on disease-
719 related outcomes and treatment-related adverse events?
720
- 721 63. In people with rheumatic disease with ILD, what is the impact of leflunomide compared to anti-CD20
722 antibody (rituximab, ocrelizumab, obinutuzumab, ofatumumab) as first line ILD treatment on disease-
723 related outcomes and treatment-related adverse events?
724
- 725 64. In people with rheumatic disease with ILD, what is the impact of azathioprine compared to anti-CD20
726 antibody (rituximab, ocrelizumab, obinutuzumab, ofatumumab) as first line ILD treatment on disease-
727 related outcomes and treatment-related adverse events?
728
- 729 65. In people with rheumatic disease with ILD, what is the impact of cyclophosphamide compared to anti-
730 CD20 antibody (rituximab, ocrelizumab, obinutuzumab, ofatumumab) as first line ILD treatment on
731 disease-related outcomes and treatment-related adverse events?
732
- 733 66. In people with rheumatic disease with ILD, what is the impact of calcineurin inhibitors compared to anti-
734 CD20 antibody (rituximab, ocrelizumab, obinutuzumab, ofatumumab) as first line ILD treatment on
735 disease-related outcomes and treatment-related adverse events?
736
- 737 67. In people with rheumatic disease with ILD, what is the impact of TNF inhibitors compared to anti-CD20
738 antibody (rituximab, ocrelizumab, obinutuzumab, ofatumumab) as first line ILD treatment on disease-
739 related outcomes and treatment-related adverse events?
740

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

- 741 68. In people with rheumatic disease with ILD, what is the impact of IL-6 receptor antagonists (tocilizumab,
742 sarilumab) compared to anti-CD20 antibody (rituximab, ocrelizumab, obinutuzumab, ofatumumab) as
743 first line ILD treatment on disease-related outcomes and treatment-related adverse events?
744
- 745 69. In people with rheumatic disease with ILD, what is the impact of abatacept compared to anti-CD20
746 antibody (rituximab, ocrelizumab, obinutuzumab, ofatumumab) as first line ILD treatment on disease-
747 related outcomes and treatment-related adverse events?
748
- 749 70. In people with rheumatic disease with ILD, what is the impact of JAK inhibitors compared to anti-CD20
750 antibody (rituximab, ocrelizumab, obinutuzumab, ofatumumab) as first line ILD treatment on disease-
751 related outcomes and treatment-related adverse events?
752
- 753 71. In people with rheumatic disease with ILD, what is the impact of nintedanib compared to anti-CD20
754 antibody (rituximab, ocrelizumab, obinutuzumab, ofatumumab) as first line ILD treatment on disease-
755 related outcomes and treatment-related adverse events?
756
- 757 72. In people with rheumatic disease with ILD, what is the impact of pirfenidone compared to anti-CD20
758 antibody (rituximab, ocrelizumab, obinutuzumab, ofatumumab) as first line ILD treatment on disease-
759 related outcomes and treatment-related adverse events?
760
- 761 73. In people with rheumatic disease with ILD, what is the impact of IVIG compared to anti-CD20 antibody
762 (rituximab, ocrelizumab, obinutuzumab, ofatumumab) as first line ILD treatment on disease-related
763 outcomes and treatment-related adverse events?
764
- 765 74. In people with rheumatic disease with ILD, what is the impact of oral prednisone compared to anti-CD20
766 antibody (rituximab, ocrelizumab, obinutuzumab, ofatumumab) as first line ILD treatment on disease-
767 related outcomes and treatment-related adverse events?
768
- 769 75. In people with rheumatic disease with ILD, what is the impact of intravenous methylprednisolone
770 compared to anti-CD20 antibody (rituximab, ocrelizumab, obinutuzumab, ofatumumab) as first line ILD
771 treatment on disease-related outcomes and treatment-related adverse events?
772
- 773 76. In people with rheumatic disease with ILD, what is the impact of plasma exchange compared to anti-
774 CD20 antibody (rituximab, ocrelizumab, obinutuzumab, ofatumumab) as first line ILD treatment on
775 disease-related outcomes and treatment-related adverse events?
776

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

- 777 77. In people with rheumatic disease with ILD, what is the impact of methotrexate compared to
778 azathioprine as first line ILD treatment on disease-related outcomes and treatment-related adverse
779 events?
780
- 781 78. In people with rheumatic disease with ILD, what is the impact of leflunomide compared to azathioprine
782 as first line ILD treatment on disease-related outcomes and treatment-related adverse events?
783
- 784 79. In people with rheumatic disease with ILD, what is the impact of cyclophosphamide compared to
785 azathioprine as first line ILD treatment on disease-related outcomes and treatment-related adverse
786 events?
787
- 788 80. In people with rheumatic disease with ILD, what is the impact of calcineurin inhibitors compared to
789 azathioprine as first line ILD treatment on disease-related outcomes and treatment-related adverse
790 events?
791
- 792 81. In people with rheumatic disease with ILD, what is the impact of TNF inhibitors compared to
793 azathioprine as first line ILD treatment on disease-related outcomes and treatment-related adverse
794 events?
795
- 796 82. In people with rheumatic disease with ILD, what is the impact of IL-6 receptor antagonists
797 (tocilizumab, sarilumab) compared to azathioprine as first line ILD treatment on disease-related
798 outcomes and treatment-related adverse events?
799
- 800 83. In people with rheumatic disease with ILD, what is the impact of abatacept compared to azathioprine as
801 first line ILD treatment on disease-related outcomes and treatment-related adverse events?
802
- 803 84. In people with rheumatic disease with ILD, what is the impact of JAK inhibitors compared to
804 azathioprine as first line ILD treatment on disease-related outcomes and treatment-related adverse
805 events?
806
- 807 85. In people with rheumatic disease with ILD, what is the impact of nintedanib compared to azathioprine as
808 first line ILD treatment on disease-related outcomes and treatment-related adverse events?
809
- 810 86. In people with rheumatic disease with ILD, what is the impact of pirfenidone compared to azathioprine
811 as first line ILD treatment on disease-related outcomes and treatment-related adverse events?
812

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

- 813 87. In people with rheumatic disease with ILD, what is the impact of IVIG compared to azathioprine as first
814 line ILD treatment on disease-related outcomes and treatment-related adverse events?
815
- 816 88. In people with rheumatic disease with ILD, what is the impact of oral prednisone compared to
817 azathioprine as first line ILD treatment on disease-related outcomes and treatment-related adverse
818 events?
819
- 820 89. In people with rheumatic disease with ILD, what is the impact of intravenous methylprednisolone
821 compared to azathioprine as first line ILD treatment on disease-related outcomes and treatment-
822 related adverse events?
823
- 824 90. In people with rheumatic disease with ILD, what is the impact of plasma exchange compared to
825 azathioprine as first line ILD treatment on disease-related outcomes and treatment-related adverse
826 events?
827
- 828 91. In people with rheumatic disease with ILD, what is the impact of methotrexate compared to
829 cyclophosphamide as first line ILD treatment on disease-related outcomes and treatment-related
830 adverse events?
831
- 832 92. In people with rheumatic disease with ILD, what is the impact of leflunomide compared to
833 cyclophosphamide as first line ILD treatment on disease-related outcomes and treatment-related
834 adverse events?
835
- 836 93. In people with rheumatic disease with ILD, what is the impact of calcineurin inhibitors compared to
837 cyclophosphamide as first line ILD treatment on disease-related outcomes and treatment-related
838 adverse events?
839
- 840 94. In people with rheumatic disease with ILD, what is the impact of TNF inhibitors compared to
841 cyclophosphamide as first line ILD treatment on disease-related outcomes and treatment-related
842 adverse events?
843
- 844 95. In people with rheumatic disease with ILD, what is the impact of IL-6 receptor antagonists (tocilizumab,
845 sarilumab) compared to cyclophosphamide as first line ILD treatment on disease-related outcomes and
846 treatment-related adverse events?
847
- 848 96. In people with rheumatic disease with ILD, what is the impact of abatacept compared to
849 cyclophosphamide as first line ILD treatment on disease-related outcomes and treatment-related

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

850 adverse events?

851

852 97. In people with rheumatic disease with ILD, what is the impact of JAK inhibitors compared to
853 cyclophosphamide as first line ILD treatment on disease-related outcomes and treatment-related
854 adverse events?

855

856 98. In people with rheumatic disease with ILD, what is the impact of nintedinib compared to
857 cyclophosphamide as first line ILD treatment on disease-related outcomes and treatment-related
858 adverse events?

859

860 99. In people with rheumatic disease with ILD, what is the impact of pirfenidone compared to
861 cyclophosphamide as first line ILD treatment on disease-related outcomes and treatment-related
862 adverse events?

863

864 100. In people with rheumatic disease with ILD, what is the impact of IVIG compared to cyclophosphamide as
865 first line ILD treatment on disease-related outcomes and treatment-related adverse events?

866

867 101. In people with rheumatic disease with ILD, what is the impact of oral prednisone compared to
868 cyclophosphamide as first line ILD treatment on disease-related outcomes and treatment-related
869 adverse events?

870

871 102. In people with rheumatic disease with ILD, what is the impact of intravenous methylprednisolone
872 compared to cyclophosphamide as first line ILD treatment on disease-related outcomes and treatment-
873 related adverse events?

874

875 103. In people with rheumatic disease with ILD, what is the impact of plasma exchange compared to
876 cyclophosphamide as first line ILD treatment on disease-related outcomes and treatment-related
877 adverse events?

878

879 104. In people with rheumatic disease with ILD, what is the impact of nintedanib compared to IL-6 receptor
880 antagonists (tocilizumab, sarilumab) as first line ILD treatment on disease-related outcomes and
881 treatment-related adverse events?

882

883 105. In people with rheumatic disease with ILD, what is the impact of referral for stem cell transplant
884 compared to optimal medical management as first line ILD treatment on disease-related outcomes and
885 treatment-related adverse events?

886

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

887 106. In people with rheumatic disease with ILD, what is the impact of referral for lung transplant compared
888 to optimal medical management as first line ILD treatment on disease-related outcomes and treatment-
889 related adverse events?

890

891 **Rheumatic disease with ILD progression after any 1st ILD therapy**

892

893 107. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
894 adding the combination of nintedanib and mycophenolate compared to adding mycophenolate alone
895 on disease-related outcomes and treatment-related adverse events?

896

897 108. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
898 adding the combination of pirfenidone and mycophenolate compared to adding mycophenolate alone
899 on disease-related outcomes and treatment-related adverse events?

900

901 109. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
902 adding methotrexate compared to adding mycophenolate on disease-related outcomes and treatment-
903 related adverse events?

904

905 110. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
906 adding leflunomide compared to adding mycophenolate on disease-related outcomes and treatment-
907 related adverse events?

908

909 111. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
910 adding azathioprine compared to adding mycophenolate on disease-related outcomes and treatment-
911 related adverse events?

912

913 112. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
914 adding cyclophosphamide compared to adding mycophenolate on disease-related outcomes and
915 treatment-related adverse events?

916

917 113. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
918 adding calcineurin inhibitors compared to adding mycophenolate on disease-related outcomes and
919 treatment-related adverse events?

920

921 114. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
922 adding TNF inhibitors compared to adding mycophenolate on disease-related outcomes and treatment-

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

- 923 related adverse events?
924
- 925 115. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
926 adding IL-6 receptor antagonists (tocilizumab, sarilumab) compared to adding mycophenolate on
927 disease-related outcomes and treatment-related adverse events?
928
- 929 116. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
930 adding anti-CD20 antibody (rituximab, ocrelizumab, obinutuzumab, ofatumumab) compared to adding
931 mycophenolate on disease-related outcomes and treatment-related adverse events?
932
- 933 117. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
934 adding abatacept compared to adding mycophenolate on disease-related outcomes and treatment-
935 related adverse events?
936
- 937 118. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
938 adding JAK inhibitors compared to adding mycophenolate on disease-related outcomes and treatment-
939 related adverse events?
940
- 941 119. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
942 adding nintedanib compared to adding mycophenolate on disease-related outcomes and treatment-
943 related adverse events?
944
- 945 120. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
946 adding pirfenidone compared to adding mycophenolate on disease-related outcomes and treatment-
947 related adverse events?
948
- 949 121. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
950 adding IVIG compared to adding mycophenolate on disease-related outcomes and treatment-related
951 adverse events?
952
- 953 122. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
954 adding oral prednisone compared to adding mycophenolate on disease-related outcomes and
955 treatment-related adverse events?
956
- 957 123. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
958 adding intravenous methylprednisolone compared to adding mycophenolate on disease-related

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

- 959 outcomes and treatment-related adverse events?
960
- 961 124. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
962 adding plasma exchange compared to adding mycophenolate on disease-related outcomes and
963 treatment-related adverse events?
964
- 965 125. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
966 adding methotrexate compared to adding anti-CD20 antibody (rituximab, ocrelizumab, obinutuzumab,
967 ofatumumab) on disease-related outcomes and treatment-related adverse events?
968
- 969 126. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
970 adding leflunomide compared to adding anti-CD20 antibody (rituximab, ocrelizumab, obinutuzumab,
971 ofatumumab) on disease-related outcomes and treatment-related adverse events?
972
- 973 127. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
974 adding azathioprine compared to adding anti-CD20 antibody (rituximab, ocrelizumab, obinutuzumab,
975 ofatumumab) on disease-related outcomes and treatment-related adverse events?
976
- 977 128. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
978 adding cyclophosphamide compared to adding anti-CD20 antibody (rituximab, ocrelizumab,
979 obinutuzumab, ofatumumab) on disease-related outcomes and treatment-related adverse events?
980
- 981 129. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
982 adding calcineurin inhibitors compared to adding anti-CD20 antibody (rituximab, ocrelizumab,
983 obinutuzumab, ofatumumab) on disease-related outcomes and treatment-related adverse events?
984
- 985 130. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
986 adding TNF inhibitors compared to adding anti-CD20 antibody (rituximab, ocrelizumab, obinutuzumab,
987 ofatumumab) on disease-related outcomes and treatment-related adverse events?
988
- 989 131. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
990 adding IL-6 receptor antagonists (tocilizumab, sarilumab) compared to adding anti-CD20 antibody
991 (rituximab, ocrelizumab, obinutuzumab, ofatumumab) on disease-related outcomes and treatment-
992 related adverse events?
993
- 994 132. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
995 adding abatacept compared to adding anti-CD20 antibody (rituximab, ocrelizumab, obinutuzumab,

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

996 ofatumumab) on disease-related outcomes and treatment-related adverse events?
997

998 133. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
999 adding JAK inhibitors compared to adding anti-CD20 antibody (rituximab, ocrelizumab, obinutuzumab,
1000 ofatumumab) on disease-related outcomes and treatment-related adverse events?
1001

1002 134. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
1003 adding nintedinib compared to adding anti-CD20 antibody (rituximab, ocrelizumab, obinutuzumab,
1004 ofatumumab) on disease-related outcomes and treatment-related adverse events?
1005

1006 135. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
1007 adding pirfenidone compared to adding anti-CD20 antibody (rituximab, ocrelizumab, obinutuzumab,
1008 ofatumumab) on disease-related outcomes and treatment-related adverse events?
1009

1010 136. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
1011 adding IVIG compared to adding anti-CD20 antibody (rituximab, ocrelizumab, obinutuzumab,
1012 ofatumumab) on disease-related outcomes and treatment-related adverse events?
1013

1014 137. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
1015 adding oral prednisone compared to adding anti-CD20 antibody (rituximab, ocrelizumab,
1016 obinutuzumab, ofatumumab) on disease-related outcomes and treatment-related adverse events?
1017

1018 138. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
1019 adding intravenous methylprednisolone compared to adding anti-CD20 antibody (rituximab,
1020 ocrelizumab, obinutuzumab, ofatumumab) on disease-related outcomes and treatment-related adverse
1021 events?
1022

1023 139. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
1024 adding plasma exchange compared to adding anti-CD20 antibody (rituximab, ocrelizumab,
1025 obinutuzumab, ofatumumab) on disease-related outcomes and treatment-related adverse events?
1026

1027 140. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
1028 adding methotrexate compared to adding azathioprine on disease-related outcomes and treatment-
1029 related adverse events?
1030

1031 141. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
1032 adding leflunomide compared to adding azathioprine on disease-related outcomes and treatment-

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

1033 related adverse events?

1034

1035 142. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
1036 adding cyclophosphamide compared to adding azathioprine on disease-related outcomes and
1037 treatment-related adverse events?

1038

1039 143. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
1040 adding calcineurin inhibitors compared to adding azathioprine on disease-related outcomes and
1041 treatment-related adverse events?

1042

1043 144. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
1044 adding TNF inhibitors compared to adding azathioprine on disease-related outcomes and treatment-
1045 related adverse events?

1046

1047 145. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
1048 adding IL-6 receptor antagonists (tocilizumab, sarilumab) compared to adding azathioprine on disease-
1049 related outcomes and treatment-related adverse events?

1050

1051 146. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
1052 adding abatacept compared to adding azathioprine on disease-related outcomes and treatment-related
1053 adverse events?

1054

1055 147. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
1056 adding JAK inhibitors compared to adding azathioprine on disease-related outcomes and treatment-
1057 related adverse events?

1058

1059 148. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
1060 adding nintedanib compared to adding azathioprine on disease-related outcomes and treatment-related
1061 adverse events?

1062

1063 149. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
1064 adding pirfenidone compared to adding azathioprine on disease-related outcomes and treatment-
1065 related adverse events?

1066

1067 150. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
1068 adding IVIG compared to adding azathioprine on disease-related outcomes and treatment-related

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

1069 adverse events?
1070

1071 151. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
1072 adding oral prednisone compared to adding azathioprine on disease-related outcomes and treatment-
1073 related adverse events?
1074

1075 152. In people with rheumatic disease ILD progression after 1st ILD therapy, what is the impact of adding
1076 intravenous methylprednisolone compared to adding azathioprine on disease-related outcomes and
1077 treatment-related adverse events?
1078

1079 153. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
1080 adding plasma exchange compared to adding azathioprine on disease-related outcomes and treatment-
1081 related adverse events?
1082

1083 154. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
1084 adding methotrexate compared to adding cyclophosphamide on disease-related outcomes and
1085 treatment-related adverse events?
1086

1087 155. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
1088 adding leflunomide compared to adding cyclophosphamide on disease-related outcomes and
1089 treatment-related adverse events?
1090

1091 156. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
1092 adding calcineurin inhibitors compared to adding cyclophosphamide on disease-related outcomes and
1093 treatment-related adverse events?
1094

1095 157. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
1096 adding TNF inhibitors compared to adding cyclophosphamide on disease-related outcomes and
1097 treatment-related adverse events?
1098

1099 158. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
1100 adding IL-6 receptor antagonists (tocilizumab, sarilumab) compared to adding cyclophosphamide on
1101 disease-related outcomes and treatment-related adverse events?
1102

1103 159. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
1104 adding abatacept compared to adding cyclophosphamide on disease-related outcomes and treatment-

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

1105 related adverse events?
1106

1107 160. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
1108 adding JAK inhibitors compared to adding cyclophosphamide on disease-related outcomes and
1109 treatment-related adverse events?
1110

1111 161. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
1112 adding nintedinib compared to adding cyclophosphamide on disease-related outcomes and treatment-
1113 related adverse events?
1114

1115 162. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
1116 adding pirfenidone compared to adding cyclophosphamide on disease-related outcomes and
1117 treatment-related adverse events?
1118

1119 163. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
1120 adding IVIG compared to adding cyclophosphamide on disease-related outcomes and treatment-related
1121 adverse events?
1122

1123 164. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
1124 adding oral prednisone compared to adding cyclophosphamide on disease-related outcomes and
1125 treatment-related adverse events?
1126

1127 165. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
1128 adding intravenous methylprednisolone compared to adding cyclophosphamide on disease-related
1129 outcomes and treatment-related adverse events?
1130

1131 166. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
1132 adding plasma exchange compared to adding cyclophosphamide on disease-related outcomes and
1133 treatment-related adverse events?
1134

1135 167. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
1136 referral for stem cell transplant compared to optimal medical management on disease-related
1137 outcomes and treatment-related adverse events?
1138

1139 168. In people with rheumatic disease with ILD progression after 1st ILD therapy, what is the impact of
1140 referral for lung transplant compared to optimal medical management on disease-related outcomes
1141 and treatment-related adverse events?

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

1142

1143 Rheumatic disease with rapidly progressive ILD

- 1144 169. In people with rheumatic disease with rapidly progressive ILD, what is the impact of daily oral
1145 prednisone compared to no daily oral prednisone as first line rapidly progressive ILD treatment on
1146 disease-related outcomes and treatment-related adverse events?
1147
- 1148 170. In people with rheumatic disease with rapidly progressive ILD, what is the impact of pulse intravenous
1149 glucocorticoids compared to no pulse intravenous glucocorticoids as first line rapidly progressive ILD
1150 treatment on disease-related outcomes and treatment-related adverse events?
1151
- 1152 171. In people with rheumatic disease with rapidly progressive ILD, what is the impact of nintedanib
1153 compared to no nintedanib as first line rapidly progressive ILD treatment on disease-related outcomes
1154 and treatment-related adverse events?
1155
- 1156 172. In people with rheumatic disease with rapidly progressive ILD, what is the impact of pirfenidone
1157 compared to no pirfenidone as first line rapidly progressive ILD treatment on disease-related outcomes
1158 and treatment-related adverse events?
1159
- 1160 173. In people with rheumatic disease with rapidly progressive ILD, what is the impact of adding nintedanib
1161 to mycophenolate compared to not adding nintedanib to mycophenolate as first line rapidly progressive
1162 ILD treatment on disease-related outcomes and treatment-related adverse events?
1163
- 1164 174. In people with rheumatic disease with rapidly progressive ILD, what is the impact of adding pirfenidone
1165 to mycophenolate compared to not adding pirfenidone to mycophenolate as first line rapidly
1166 progressive ILD treatment on disease-related outcomes and treatment-related adverse events?
1167
- 1168 175. In people with rheumatic disease with rapidly progressive ILD, what is the impact of upfront
1169 combination of nintedanib with mycophenolate compared to mycophenolate alone as first line rapidly
1170 progressive ILD treatment on disease-related outcomes and treatment-related adverse events?
1171
- 1172 176. In people with rheumatic disease with rapidly progressive ILD, what is the impact of upfront
1173 combination of pirfenidone with mycophenolate compared to mycophenolate alone as first line rapidly
1174 progressive ILD treatment on disease-related outcomes and treatment-related adverse events?
1175
- 1176 177. In people with rheumatic disease with rapidly progressive ILD, what is the impact of methotrexate
1177 compared to mycophenolate as first line rapidly progressive ILD treatment on disease-related outcomes

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

1178 and treatment-related adverse events?

1179

1180 178. In people with rheumatic disease with rapidly progressive ILD, what is the impact of leflunomide
1181 compared to mycophenolate as first line rapidly progressive ILD treatment on disease-related outcomes
1182 and treatment-related adverse events?

1183

1184 179. In people with rheumatic disease with rapidly progressive ILD, what is the impact of azathioprine
1185 compared to mycophenolate as first line rapidly progressive ILD treatment on disease-related outcomes
1186 and treatment-related adverse events?

1187

1188 180. In people with rheumatic disease with rapidly progressive ILD, what is the impact of cyclophosphamide
1189 compared to mycophenolate as first line rapidly progressive ILD treatment on disease-related outcomes
1190 and treatment-related adverse events?

1191

1192 181. In people with rheumatic disease with rapidly progressive ILD, what is the impact of calcineurin
1193 inhibitors compared to mycophenolate as first line rapidly progressive ILD treatment on disease-related
1194 outcomes and treatment-related adverse events?

1195

1196 182. In people with rheumatic disease with rapidly progressive ILD, what is the impact of TNF inhibitors
1197 compared to mycophenolate as first line rapidly progressive ILD treatment on disease-related outcomes
1198 and treatment-related adverse events?

1199

1200 183. In people with rheumatic disease with rapidly progressive ILD, what is the impact of IL-6 receptor
1201 antagonists (tocilizumab, sarilumab) compared to mycophenolate as first line rapidly progressive ILD
1202 treatment on disease-related outcomes and treatment-related adverse events?

1203

1204 184. In people with rheumatic disease with rapidly progressive ILD, what is the impact of anti-CD20 antibody
1205 (rituximab, ocrelizumab, obinutuzumab, ofatumumab) compared to mycophenolate as first line rapidly
1206 progressive ILD treatment on disease-related outcomes and treatment-related adverse events?

1207

1208 185. In people with rheumatic disease with rapidly progressive ILD, what is the impact of abatacept
1209 compared to mycophenolate as first line rapidly progressive ILD treatment on disease-related outcomes
1210 and treatment-related adverse events?

1211

1212 186. In people with rheumatic disease with rapidly progressive ILD, what is the impact of JAK inhibitors
1213 compared to mycophenolate as first line rapidly progressive ILD treatment on disease-related outcomes

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

1214 and treatment-related adverse events?

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1216 187. In people with rheumatic disease with rapidly progressive ILD, what is the impact of nintedinib
1217 compared to mycophenolate as first line rapidly progressive ILD treatment on disease-related outcomes
1218 and treatment-related adverse events?

1219

1220 188. In people with rheumatic disease with rapidly progressive ILD, what is the impact of pirfenidone
1221 compared to mycophenolate as first line rapidly progressive ILD treatment on disease-related outcomes
1222 and treatment-related adverse events?

1223

1224 189. In people with rheumatic disease with rapidly progressive ILD, what is the impact of IVIG compared to
1225 mycophenolate as first line rapidly progressive ILD treatment on disease-related outcomes and
1226 treatment-related adverse events?

1227

1228 190. In people with rheumatic disease with rapidly progressive ILD, what is the impact of oral prednisone
1229 compared to mycophenolate as first line rapidly progressive ILD treatment on disease-related outcomes
1230 and treatment-related adverse events?

1231

1232 191. In people with rheumatic disease with rapidly progressive ILD, what is the impact of intravenous
1233 methylprednisolone compared to mycophenolate as first line rapidly progressive ILD treatment on
1234 disease-related outcomes and treatment-related adverse events?

1235

1236 192. In people with rheumatic disease with rapidly progressive ILD, what is the impact of plasma exchange
1237 compared to mycophenolate as first line rapidly progressive ILD treatment on disease-related outcomes
1238 and treatment-related adverse events?

1239

1240 193. In people with rheumatic disease with rapidly progressive ILD, what is the impact of methotrexate
1241 compared to anti-CD20 antibody (rituximab, ocrelizumab, obinutuzumab, ofatumumab) as first line
1242 rapidly progressive ILD treatment on disease-related outcomes and treatment-related adverse events?

1243

1244 194. In people with rheumatic disease with rapidly progressive ILD, what is the impact of leflunomide
1245 compared to anti-CD20 antibody (rituximab, ocrelizumab, obinutuzumab, ofatumumab) as first line
1246 rapidly progressive ILD treatment on disease-related outcomes and treatment-related adverse events?

1247

1248 195. In people with rheumatic disease with rapidly progressive ILD, what is the impact of azathioprine
1249 compared to anti-CD20 antibody (rituximab, ocrelizumab, obinutuzumab, ofatumumab) as first line

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

- 1250 rapidly progressive ILD treatment on disease-related outcomes and treatment-related adverse events?
1251
- 1252 196. In people with rheumatic disease with rapidly progressive ILD, what is the impact of cyclophosphamide
1253 compared to anti-CD20 antibody (rituximab, ocrelizumab, obinutuzumab, ofatumumab) as first line
1254 rapidly progressive ILD treatment on disease-related outcomes and treatment-related adverse events?
1255
- 1256 197. In people with rheumatic disease with rapidly progressive ILD, what is the impact of calcineurin
1257 inhibitors compared to anti-CD20 (rituximab, ocrelizumab, obinutuzumab, ofatumumab) antibody as
1258 first line rapidly progressive ILD treatment on disease-related outcomes and treatment-related adverse
1259 events?
1260
- 1261 198. In people with rheumatic disease with rapidly progressive ILD, what is the impact of TNF inhibitors
1262 compared to anti-CD20 antibody (rituximab, ocrelizumab, obinutuzumab, ofatumumab) as first line
1263 rapidly progressive ILD treatment on disease-related outcomes and treatment-related adverse events?
1264
- 1265 199. In people with rheumatic disease with rapidly progressive ILD, what is the impact of IL-6 receptor
1266 antagonists (tocilizumab, sarilumab) compared to anti-CD20 antibody (rituximab, ocrelizumab,
1267 obinutuzumab, ofatumumab) as first line rapidly progressive ILD treatment on disease-related
1268 outcomes and treatment-related adverse events?
1269
- 1270 200. In people with rheumatic disease with rapidly progressive ILD, what is the impact of abatacept
1271 compared to anti-CD20 antibody (rituximab, ocrelizumab, obinutuzumab, ofatumumab) as first line
1272 rapidly progressive ILD treatment on disease-related outcomes and treatment-related adverse events?
1273
- 1274 201. In people with rheumatic disease with rapidly progressive ILD, what is the impact of JAK inhibitors
1275 compared to anti-CD20 antibody (rituximab, ocrelizumab, obinutuzumab, ofatumumab) as first line
1276 rapidly progressive ILD treatment on disease-related outcomes and treatment-related adverse events?
1277
- 1278 202. In people with rheumatic disease with rapidly progressive ILD, what is the impact of nintedinib
1279 compared to anti-CD20 antibody (rituximab, ocrelizumab, obinutuzumab, ofatumumab) as first line
1280 rapidly progressive ILD treatment on disease-related outcomes and treatment-related adverse events?
1281
- 1282 203. In people with rheumatic disease with rapidly progressive ILD, what is the impact of pirfenidone
1283 compared to anti-CD20 antibody (rituximab, ocrelizumab, obinutuzumab, ofatumumab) as first line
1284 rapidly progressive ILD treatment on disease-related outcomes and treatment-related adverse events?
1285

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

- 1286 204. In people with rheumatic disease with rapidly progressive ILD, what is the impact of IVIG compared to
1287 anti-CD20 antibody (rituximab, ocrelizumab, obinutuzumab, ofatumumab) as first line rapidly
1288 progressive ILD treatment on disease-related outcomes and treatment-related adverse events?
1289
- 1290 205. In people with rheumatic disease with rapidly progressive ILD, what is the impact of oral prednisone
1291 compared to anti-CD20 antibody (rituximab, ocrelizumab, obinutuzumab, ofatumumab) as first line
1292 rapidly progressive ILD treatment on disease-related outcomes and treatment-related adverse events?
1293
- 1294 206. In people with rheumatic disease with rapidly progressive ILD, what is the impact of intravenous
1295 methylprednisolone compared to anti-CD20 antibody (rituximab, ocrelizumab, obinutuzumab,
1296 ofatumumab) as first line rapidly progressive ILD treatment on disease-related outcomes and
1297 treatment-related adverse events?
1298
- 1299 207. In people with rheumatic disease with rapidly progressive ILD, what is the impact of plasma exchange
1300 compared to anti-CD20 antibody (rituximab, ocrelizumab, obinutuzumab, ofatumumab) as first line
1301 rapidly progressive ILD treatment on disease-related outcomes and treatment-related adverse events?
1302
- 1303 208. In people with rheumatic disease with rapidly progressive ILD, what is the impact of methotrexate
1304 compared to azathioprine as first line rapidly progressive ILD treatment on disease-related outcomes
1305 and treatment-related adverse events?
1306
- 1307 209. In people with rheumatic disease with rapidly progressive ILD, what is the impact of leflunomide
1308 compared to azathioprine as first line rapidly progressive ILD treatment on disease-related outcomes
1309 and treatment-related adverse events?
1310
- 1311 210. In people with rheumatic disease with rapidly progressive ILD, what is the impact of cyclophosphamide
1312 compared to azathioprine as first line rapidly progressive ILD treatment on disease-related outcomes
1313 and treatment-related adverse events?
1314
- 1315 211. In people with rheumatic disease with rapidly progressive ILD, what is the impact of calcineurin
1316 inhibitors compared to azathioprine as first line rapidly progressive ILD treatment on disease-related
1317 outcomes and treatment-related adverse events?
1318
- 1319 212. In people with rheumatic disease with rapidly progressive ILD, what is the impact of TNF inhibitors
1320 compared to azathioprine as first line rapidly progressive ILD treatment on disease-related outcomes
1321 and treatment-related adverse events?
1322

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

- 1323 213. In people with rheumatic disease with rapidly progressive ILD, what is the impact of IL-6 receptor
1324 antagonists (tocilizumab, sarilumab) compared to azathioprine as first line rapidly progressive ILD
1325 treatment on disease-related outcomes and treatment-related adverse events?
1326
- 1327 214. In people with rheumatic disease with rapidly progressive ILD, what is the impact of abatacept
1328 compared to azathioprine as first line rapidly progressive ILD treatment on disease-related outcomes
1329 and treatment-related adverse events?
1330
- 1331 215. In people with rheumatic disease with rapidly progressive ILD, what is the impact of JAK inhibitors
1332 compared to azathioprine as first line rapidly progressive ILD treatment on disease-related outcomes
1333 and treatment-related adverse events?
1334
- 1335 216. In people with rheumatic disease with rapidly progressive ILD, what is the impact of nintedanib
1336 compared to azathioprine as first line rapidly progressive ILD treatment on disease-related outcomes
1337 and treatment-related adverse events?
1338
- 1339 217. In people with rheumatic disease with rapidly progressive ILD, what is the impact of pirfenidone
1340 compared to azathioprine as first line rapidly progressive ILD treatment on disease-related outcomes
1341 and treatment-related adverse events?
1342
- 1343 218. In people with rheumatic disease with rapidly progressive ILD, what is the impact of IVIG compared to
1344 azathioprine as first line rapidly progressive ILD treatment on disease-related outcomes and treatment-
1345 related adverse events?
1346
- 1347 219. In people with rheumatic disease with rapidly progressive ILD, what is the impact of oral prednisone
1348 compared to azathioprine as first line rapidly progressive ILD treatment on disease-related outcomes
1349 and treatment-related adverse events?
1350
- 1351 220. In people with rheumatic disease with rapidly progressive ILD, what is the impact of intravenous
1352 methylprednisolone compared to azathioprine as first line rapidly progressive ILD treatment on disease-
1353 related outcomes and treatment-related adverse events?
1354
- 1355 221. In people with rheumatic disease with rapidly progressive ILD, what is the impact of plasma exchange
1356 compared to azathioprine as first line rapidly progressive ILD treatment on disease-related outcomes
1357 and treatment-related adverse events?
1358

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

- 1359 222. In people with rheumatic disease with rapidly progressive ILD, what is the impact of methotrexate
1360 compared to cyclophosphamide as first line rapidly progressive ILD treatment on disease-related
1361 outcomes and treatment-related adverse events?
1362
- 1363 223. In people with rheumatic disease with rapidly progressive ILD, what is the impact of leflunomide
1364 compared to cyclophosphamide as first line rapidly progressive ILD treatment on disease-related
1365 outcomes and treatment-related adverse events?
1366
- 1367 224. In people with rheumatic disease with rapidly progressive ILD, what is the impact of calcineurin
1368 inhibitors compared to cyclophosphamide as first line rapidly progressive ILD treatment on disease-
1369 related outcomes and treatment-related adverse events?
1370
- 1371 225. In people with rheumatic disease with rapidly progressive ILD, what is the impact of TNF inhibitors
1372 compared to cyclophosphamide as first line rapidly progressive ILD treatment on disease-related
1373 outcomes and treatment-related adverse events?
1374
- 1375 226. In people with rheumatic disease with rapidly progressive ILD, what is the impact of IL-6 receptor
1376 antagonists (tocilizumab, sarilumab) compared to cyclophosphamide as first line rapidly progressive ILD
1377 treatment on disease-related outcomes and treatment-related adverse events?
1378
- 1379 227. In people with rheumatic disease with rapidly progressive ILD, what is the impact of abatacept
1380 compared to cyclophosphamide as first line rapidly progressive ILD treatment on disease-related
1381 outcomes and treatment-related adverse events?
1382
- 1383 228. In people with rheumatic disease with rapidly progressive ILD, what is the impact of JAK inhibitors
1384 compared to cyclophosphamide as first line rapidly progressive ILD treatment on disease-related
1385 outcomes and treatment-related adverse events?
1386
- 1387 229. In people with rheumatic disease with rapidly progressive ILD, what is the impact of nintedinib
1388 compared to cyclophosphamide as first line rapidly progressive ILD treatment on disease-related
1389 outcomes and treatment-related adverse events?
1390
- 1391 230. In people with rheumatic disease with rapidly progressive ILD, what is the impact of pirfenidone
1392 compared to cyclophosphamide as first line rapidly progressive ILD treatment on disease-related
1393 outcomes and treatment-related adverse events?
1394

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

- 1395 231. In people with rheumatic disease with rapidly progressive ILD, what is the impact of IVIG compared to
1396 cyclophosphamide as first line rapidly progressive ILD treatment on disease-related outcomes and
1397 treatment-related adverse events?
1398
- 1399 232. In people with rheumatic disease with rapidly progressive ILD, what is the impact of oral prednisone
1400 compared to cyclophosphamide as first line rapidly progressive ILD treatment on disease-related
1401 outcomes and treatment-related adverse events?
1402
- 1403 233. In people with rheumatic disease with rapidly progressive ILD, what is the impact of intravenous
1404 methylprednisolone compared to cyclophosphamide as first line rapidly progressive ILD treatment on
1405 disease-related outcomes and treatment-related adverse events?
1406
- 1407 234. In people with rheumatic disease with rapidly progressive ILD, what is the impact of plasma exchange
1408 compared to cyclophosphamide as first line rapidly progressive ILD treatment on disease-related
1409 outcomes and treatment-related adverse events?
1410
- 1411 235. In people with rheumatic disease with rapidly progressive ILD, what is the impact of dual combination
1412 therapy* compared to monotherapy† as first line rapidly progressive ILD treatment on disease-related
1413 outcomes and treatment-related adverse events?
1414
- 1415 236. In people with rheumatic disease with rapidly progressive ILD, what is the impact of triple combination
1416 therapy‡ compared to monotherapy† as first line rapidly progressive ILD treatment on disease-related
1417 outcomes and treatment-related adverse events?
1418
- 1419 237. In people with rheumatic disease with rapidly progressive ILD, what is the impact of triple combination
1420 therapy‡ compared to dual combination therapy* as first line rapidly progressive ILD treatment on
1421 disease-related outcomes and treatment-related adverse events?
1422
- 1423 238. In people with rheumatic disease with rapidly progressive ILD, what is the impact of using IVIG and/or
1424 plasma exchange in addition to monotherapy†, dual combination therapy*, or triple combination
1425 therapy‡ compared to using monotherapy†, dual combination therapy*, or triple combination therapy‡
1426 alone as first line rapidly progressive ILD treatment on disease-related outcomes and treatment-related
1427 adverse events?
1428
- 1429 239. In people with rheumatic disease with rapidly progressive ILD, what is the impact of using an antifibrotic
1430 (e.g., nintedanib or pirfenidone) in addition to monotherapy†, dual combination therapy*, or triple
1431 combination therapy‡ compared to using monotherapy†, dual combination therapy*, or triple

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

- 1432 combination therapy‡ alone as first line rapidly progressive ILD treatment on disease-related outcomes
1433 and treatment-related adverse events?
1434
- 1435 240. In people with rheumatic disease with rapidly progressive ILD, what is the impact of referral for stem
1436 cell transplant compared to optimal medical management as first line rapidly progressive ILD treatment
1437 on disease-related outcomes and treatment-related adverse events?
1438
- 1439 241. In people with rheumatic disease with rapidly progressive ILD, what is the impact of referral for lung
1440 transplant compared to optimal medical management as first line rapidly progressive ILD treatment on
1441 disease-related outcomes and treatment-related adverse events?
1442
- 1443 † Monotherapy examples: oral prednisone/intravenous methylprednisolone, or mycophenolate, or
1444 azathioprine, or a calcineurin inhibitor, or rituximab, or cyclophosphamide
- 1445 * Dual combination therapy examples: oral prednisone/intravenous methylprednisolone and mycophenolate,
1446 or oral prednisone/intravenous methylprednisolone and azathioprine, or oral prednisone/intravenous
1447 methylprednisolone and a calcineurin inhibitor, or oral prednisone/intravenous methylprednisolone and
1448 rituximab, or oral prednisone/intravenous methylprednisolone and cyclophosphamide, or oral
1449 prednisone/intravenous methylprednisolone and a JAK inhibitor
- 1450 ‡ Triple combination therapy examples: oral prednisone/intravenous methylprednisolone and rituximab and
1451 cyclophosphamide, or oral prednisone/intravenous methylprednisolone and cyclophosphamide and a
1452 calcineurin inhibitor, or oral prednisone/intravenous methylprednisolone and mycophenolate and a
1453 calcineurin inhibitor, or oral prednisone/intravenous methylprednisolone and mycophenolate and abatacept,
1454 or oral prednisone/intravenous methylprednisolone and rituximab and mycophenolate

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

APPENDIX B – INCLUSION/EXCLUSION CRITERIA

POPULATIONS

Include

- Adults
- Rheumatoid arthritis, Systemic sclerosis (Scleroderma), Mixed Connective Tissue Disease (MCTD), Polymyositis, Dermatomyositis, MDA5 Dermatomyositis, Immune Mediated Necrotizing Myositis, Antisynthetase syndrome, Sjogren’s syndrome
- Progressive Fibrosing ILD

Exclude

- Pediatrics (age 16 or younger)
- Juvenile scleroderma, juvenile systemic sclerosis, juvenile dermatomyositis, juvenile idiopathic arthritis
- Sarcoidosis, Interstitial Pneumonia with Autoimmune Features (IPAF), ankylosing spondylitis, ANCA-associated vasculitis, Systemic lupus erythematosus, Undifferentiated connective tissue disease
- Idiopathic Pulmonary Fibrosis
- Idiopathic interstitial pneumonias
- Unclassifiable ILD
- Overlap syndromes (e.g., SSc+myositis, RA+SSc, et)

INTERVENTIONS

Include

- Pulmonary Function Tests (PFTs)
- History/physical alone (e.g., shortness of breath (dyspnea), functional class and physician examination: crackles on auscultation)
- High resolution CT Thorax
- 6-minute walk test distance
- Ambulatory desaturation
- Chest radiograph (chest x-ray)
- Bronchoscopy (may include broncho-alveolar lavage, transbronchial biopsy)
- Surgical lung biopsy
- csDMARDs: methotrexate, leflunomide, azathioprine, cyclophosphamide, mycophenolate, calcineurin inhibitors (tacrolimus, cyclosporine)
- bDMARDs: TNF inhibitors (etanercept, adalimumab, infliximab, golimumab, certolizumab pegol), IL-6 receptor antagonists (tocilizumab, sarilumab), anti-CD20 antibody (rituximab, ocrelizumab, obinutuzumab, ofatumumab), abatacept

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
 Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

- 1493 • tsDMARDs: JAK inhibitors (tofacitinib, baricitinib, upadacitinib)
- 1494 • Others: Oral prednisone, intravenous methylprednisolone, intravenous immunoglobulin (IVIG), plasma
- 1495 exchange (plasmapheresis)
- 1496 • Antifibrotics: Pirfenidone, Nintedanib
- 1497 • Stem cell transplant (autologous, mesenchymal stem cells, hematopoietic, myeloablative, non-myeloablative)
- 1498 • Lung Transplant
- 1499
- 1500 **Exclude**
- 1501 • Vaccines: influenza, COVID-19, MMR, pneumococcus vaccine (refer to 2022 ACR vaccine guideline, presently in
- 1502 journal review; summary is [online](#))
- 1503 • Education (self-management of oxygen, ILD disease) (*will mention in Introduction or Discussion section*)
- 1504 • Physiotherapy (chest physiotherapy, airway clearance, incentive spirometry), Exercise (aerobic, resistance training,
- 1505 yoga, tai chi), Pulmonary Rehabilitation (cardio-pulmonary rehabilitation, resistance training, in a center versus
- 1506 home)
- 1507 • Oxygen (oxygen desaturation at rest, oxygen desaturation <88% with exercise)
- 1508 • Palliative care (cough, pain, air hunger, end stage, end of life planning, when to initiate, what to initiate)
- 1509 • Smoking cessation
- 1510 • Fundoplication
- 1511 • GI medications: proton pump inhibitors, H2 blockers, promotility agents
- 1512 • Ibritumomab (is anti-CD20, but it is radioimmunotherapy)
- 1513 • Basiliximab
- 1514
- 1515 **COMPARATORS**
- 1516 **Include**
- 1517 • No test
- 1518 • History/physical alone (e.g., shortness of breath (dyspnea), functional class and physician examination: crackles on
- 1519 auscultation)
- 1520 • High resolution CT Thorax
- 1521 • Bronchoscopy (may include broncho-alveolar lavage, transbronchial biopsy, cryobiopsy)
- 1522 • 6-minute walk test distance
- 1523 • Ambulatory desaturation
- 1524 • PFTs
- 1525 • Placebo, no treatment
- 1526 • Mycophenolate
- 1527 • antiAnti-CD20 antibody (rituximab, ocrelizumab, obinutuzumab, ofatumumab)
- 1528 • Azathioprine
- 1529 • Cyclophosphamide

**American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease**

Project Plan – August 2022

- 1530 • Stem cell transplant (autologous, mesenchymal stem cells, hematopoietic, myeloablative, non-myeloablative)
1531 • Lung transplant
1532

1533 **OUTCOMES** (see Appendix C)
1534

1535 **STUDY DESIGN** (includes only studies published in English language)

1536 For all PICO questions, we will include randomized or non-randomized controlled trials (this includes case-
1537 control studies). To capture adverse events, we will also consider open-label extension studies of RCTs or
1538 other longitudinal observational studies that focus on safety and tolerability. For PICO questions that focus on
1539 assessing the accuracy of screening tools, we will also include studies without an independent control group,
1540 specifically cohort and cross-sectional studies. We will also include existing systematic reviews and guidelines
1541 from other societies **only** to confirm that we have included all relevant reference.

1542 **Include**

- 1543 • RCTs, including:
1544 ○ Open-label extensions of RCTs with placebo involved
1545 • Non-randomized controlled studies, including
1546 ○ Case-control studies
1547 • Cohort studies
1548 • Cross-sectional studies
1549 • Longitudinal studies (focusing on safety and tolerability)
1550 • Systematic reviews and Guidelines from other societies
1551

1552 [NOTE: If there has been a recently done, well-done systematic review on the exact PICO that ACR is asking,
1553 then that systematic review could be considered for use in the guideline; primary study data would still need to
1554 be pulled in the ACR's database, though.]

1555 **Exclude**

- 1556 • Abstracts
1557 • Case reports
1558 • Narrative reviews
1559 • Prevalence studies
1560 • Economic studies, e.g., cost-effectiveness studies
1561 • Drug adherence studies
1562 • Studies of risk factors
1563 • Foreign language studies
1564 • Studies with irrelevant population, interventions, or outcomes
1565 • Animal studies

American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of
Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease

Project Plan – August 2022

1566 **APPENDIX C: OUTCOMES**

1567

1568 **Screening**

1569 **Critical outcomes:**

- 1570 • Diagnostic accuracy
- 1571 • Disease-related outcomes¹
- 1572 • Diagnostic testing-related adverse events

1573

1574 **Monitoring**

1575 **Critical outcomes:**

- 1576 • Responsiveness/sensitivity to change of the test
- 1577 • Disease-related outcomes¹
- 1578 • Treatment-related serious adverse events
- 1579 • Testing-related adverse events

1580

1581 **Medical management**

- 1582 • Disease-related outcomes¹
- 1583 • Treatment-related adverse events

1584

1585 ¹**Critical outcomes:** mortality, disability, health related quality of life, adverse events (serious adverse events,
1586 toxicity leading to discontinuation). **Surrogate outcomes:** disease activity/disease progression defined by forced
1587 vital capacity (FVC), diffusion capacity for carbon monoxide (DLCO), CT thorax: extent of disease, disease
1588 progression.

1589

1590

1591

American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease

Project Plan – August 2022

APPENDIX D: DISCLOSURES

Participant Disclosures - American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of Interstitial Lung Disease in Patients with Systemic Autoimmune Rheumatic Disease							
<p>In order for the College to most effectively further its mission and to otherwise maintain its excellent reputation in the medical community and with the public, it is important that confidence in the College’s integrity be maintained. The cornerstone of the ACR’s Disclosure Policy is disclosure of actual and potential conflicts so that they can be evaluated by the College in order to avoid undue influence of potential conflicts. The purpose of the ACR’s Disclosure Policy is identification of relationships which may pose actual or potential conflicts. These actual or potential conflicts can then be evaluated by the College so that adjustments can be made that will avoid any undue influence. This policy is based on the principle that, in many cases, full disclosure of the actual or potentially conflicting relationship will of itself suffice to protect the integrity of the College and its interests.</p>							
Participants	Role	Primary Employer	Interest Held By	Interest Type	Entity/Licensee	Additional Information	Value
Sindhu R. Johnson	Core Team - Co-PI	Toronto Western Hospital	Self	Independent Contractor - Editorial Board	Best Practice and Clinical Rheumatology		
			Self	Independent Contractor - Associate Editor	Journal of Rheumatology		
			Spouse/Partner	Independent Contractor - Editorial Board Member	Anesthesiology		
			Spouse/Partner	Independent Contractor - Editorial Board Member	Canadian Journal of Anesthesia		
			Spouse/Partner	Independent Contractor - Consultant	Edwards Lifesciences		
			Spouse/Partner	Independent Contractor - Consultant	Surgical Safety Technologies		
Elana Bernstein	Core Team - Co-PI	Columbia University/New York-Presbyterian Hospital	Self	Grant / Contract	U.S. Department of Defense		\$750,000.00
			Self	Employment	James J. Peters VA Medical Center		
			Self	Employment	Columbia University		
			Self	Grant / Contract	Scleroderma Research Foundation	Columbia receives fees from the Scleroderma Research Foundation based on the number of new patient study visits and follow-up patient	\$52,000.00

American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease

Project Plan – August 2022

						study visits conducted.	
			Self	Grant / Contract	National Institutes of Health		\$801,500.00
			Self	Independent Contractor - IC Iloprost for SSC-associated Raynaud's	Eicos	Total direct costs: \$16,358.19 per patient	
			Self	Independent Contractor - Data And Safety Monitoring	UCLA Health System		\$20,000.00
			Self	Independent Contractor - A Phase 2, Open-label, Multicenter Study to Evaluate the Efficacy and Safety of Belumosudil in dcSSc	Kadmon Pharmaceuticals LLC	Total direct costs: \$40,697 per patient	
			Self	Grant / Contract	Boehringer Ingelheim		\$500,000.00
			Self	Independent Contractor - Consultant	Boehringer Ingelheim		\$6,526.25
			Self	Grant / Contract	Pfizer		\$150,000.00
			Self	Independent Contractor - Scientific Advisory Board	Boehringer Ingelheim	I did not accept any compensation for participating in this Scientific Advisory Board.	

American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease

Project Plan – August 2022

Marcy B. Bolster	Core Team - Content expert	Massachusetts General Hospital	Self	Grant / Contract	Genentech	Clinical trial I helped with data collection but did not receive salary support from this trial. I am not privy to the contracted amount. I thought I should disclose my relationship since my name is listed as a co-investigator and co-author on the publication. Please let me know if additional information is needed.	\$1.00
			Self	Grant / Contract	Cumberland Pharmaceuticals	I am a co-investigator for this clinical trial	\$31,699.00
			Self	Independent Contractor - Consultant	Merck Sharp & Dohme Corporation		
			Self	Independent Contractor - Associate Editor	PracticeUpdate		\$10,000.00
			Self	Independent Contractor - Member, ABIM Rheumatology Longitudinal Assessment Program (LAP) Approval Committee	American Board of Internal Medicine		
			Self	Independent Contractor - Co-investigator	Corbus		
			custodial account with child who is no longer a dependent	Stock	Johnson and Johnson		\$5,000.00
			Self	Independent Contractor - Principal Investigator	Rheumatology Research Foundation	Grant recipient for salary support	
			Self	Independent Contractor - Principal Investigator	Rheumatology Research Foundation	Grant recipient for salary support	

American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease

Project Plan – August 2022

Jonathan H. Chung	Core Team - Content expert	University of Chicago				Nothing to disclose	
Sonye Danoff	Core Team - Content expert	Johns Hopkins University School of Medicine	Self	Grant / Contract	Boehringer Ingelheim	Open label extension of INBUILD clinical trial	\$15,000.00
			Self	Grant / Contract	Boehringer Ingelheim		\$500,000.00
			Self	Independent Contractor - Consultant	CSL Behring	This is a 4- hour virtual multi-disciplinary advisory board on autoimmune myositis	\$3,000.00
			Self	Grant / Contract	Bristol Myers Squibb Company	This is a clinical trial of an LPA inhibitor in Progressive-fibrosing ILD	\$125,000.00
			Self	Independent Contractor - Consultant	Boehringer Ingelheim		\$2,600.00
			Self	Independent Contractor - Data And Safety Monitoring	Galapagos		\$1,000.00
			Self	Employment	Pulmonary Fibrosis Foundation		
			Self	Fiduciary Officer	American Thoracic Society		
Michael George	Core Team - Content expert	Penn Medicine	Self	Independent Contractor - Consultant	AbbVie		\$4,085.00
			Self	Employment	Perelman School of Medicine, University of Pennsylvania		
			Self	Independent Contractor - Consultant	Global Healthy Living Foundation		
			Self	Grant / Contract	GlaxoSmithKline		\$61,000.00
			Self	Independent Contractor - Consultant	Dysimmune Diseases Foundation		\$2,600.00
			Self	Independent Contractor - Consultant	Global Healthy Living Foundation		\$3,000.00
Dinesh Khanna	Core Team - Content expert	University of Michigan	Self	Independent Contractor - Consultant	Genentech Foundation		\$4,000.00
			Self	Independent Contractor - Consultant	CSL Behring		\$1,000.00
			Self	Independent Contractor - Consultant	AbbVie		\$4,000.00
			Self	Independent Contractor - Consultant	Actelion Pharmaceuticals		\$6,000.00
			Self	Independent Contractor - Consultant	Prometheus		\$5,000.00

American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease

Project Plan – August 2022

			Self	Independent Contractor - Consultant	Horizon Pharma plc		\$20,000.00
			Self	Stock Option	Eicos Sciences	Chief Medical Officer	
			Self	Independent Contractor - Consultant	Boehringer Ingelheim		\$80,000.00
Ilya Ivlev	Core Team - Literature Review Co-Leader	ECRI Institute	Self	Grant / Contract	U.S. Department of Defense	Role-co-lead	\$0.00
			Self	Grant / Contract	Agency for Healthcare Research and Quality	Goal 1: The purpose of this Task Order is to conduct systematic evidence reviews that the Task Force will use to make new or update existing recommendations Goal 2: To synthesize available evidence to support primary care guidelines development.	\$0.00
			Self	Independent Contractor - Affiliate Investigator	Kaiser Permanente		
			Self	Grant / Contract	National Institute on Aging		\$60,000.00
			Self	Employment	ECRI		
			Self	Grant / Contract	National Cancer Institute	To synthesize available evidence to support primary care guidelines development. Role: Research Curator, evidence synthesis for actionability	\$0.00
			Self	Employment	Kaiser Permanente		
			Self	Grant / Contract	Agency for Healthcare Research and Quality		\$300,000.00

American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease

Project Plan – August 2022

Stacey Uhl	Core Team - Literature Review Co-Leader	ECRI Institute				Nothing to disclose	
Gordon Guyatt	Core Team - GRADE Expert	McMaster University				Nothing to disclose	
Reza Mirza	Core Team - GRADE/Methodological Contributor	McMaster University				Nothing to disclose	
Sandeep Agarwal	Lit Review Team	Baylor College of Medicine				Nothing to disclose	
Danielle Antin-Ozerkis	Lit Review Team	Yale School of Medicine	Self	Grant / Contract	Pliant Therapeutics	Grant paid to institution for work completed, patients enrolled	\$130,999.00
			Self	Grant / Contract	Galacto Biotech AB	Grant paid to institution for work completed, patients enrolled	\$116,975.00
			Self	Grant / Contract	Galapagos	Grant paid to institution for work completed, patients enrolled	\$181,000.00
			Self	Grant / Contract	FibroGen	Grant paid to institution for work completed, patients enrolled	\$133,000.00
			Self	Stock	Amgen		\$100,000.00
			Self	Grant / Contract	Boehringer Ingelheim	Grant paid to institution for work completed, patients enrolled	\$126,683.00
			Self	Grant / Contract	Boehringer Ingelheim	Grant paid to institution for work completed, patients enrolled	\$120,811.00
			Self	Stock	Pfizer		\$15,000.00
			Self	Stock	AbbVie		\$100,000.00

American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease

Project Plan – August 2022

			Self	Grant / Contract	Genentech	Grant paid to institution for work completed, patients enrolled	\$211,000.00
Bradford Bemiss	Lit Review Team	Loyola University Medical Center	Self	Independent Contractor - Speakers bureau	Boehringer Ingelheim		
Vaidehi Chowdhary	Lit Review Team	Yale School of Medicine	Self	Independent Contractor - International Editor	International Journal of Rheumatic Diseases		
			Self	Grant / Contract	MCIC vermont	Title: Communication and Management of Test Results in Patients Initiating Biologic Agents PI on the Grant awarded to the section. No compensation for salary, or consultation. No effort paid for self.	\$75,000.00
			Self	Independent Contractor - International editorial board member	Indian Journal of Rheumatology		
Jane E. Dematte D'Amico	Lit Review Team	Northwestern Medicine	Self	Independent Contractor - Site Principal Investigator	United Therapeutics Corporation		
			Self	Independent Contractor - Site Principle Investigator	FibroGen		
			Self	Independent Contractor - Site Prinicpal Investigator	Genentech		
			Self	Independent Contractor - Site Principal Investigator	Boehringer Ingelheim		
Robert Hallowell	Lit Review Team	Massachusetts General Hospital	Self	Independent Contractor - Consultant	Genentech		\$4,000.00
			Self	Independent Contractor - Consultant	Boehringer Ingelheim		\$2,500.00
			Self	Independent Contractor - Consultant	Boehringer Ingelheim		\$5,000.00
			Self	Medical Advisory board for the Myositis Association.			

American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease

Project Plan – August 2022

Alicia M. Hinze	Lit Review Team	Mayo Clinic	Contract with my Employer Mayo Clinic	Grant / Contract	Rheumatology Research Foundation	Career development award which supports engagement in research and career development activities to evaluate radiomic biomarkers for ILD progression in systemic sclerosis.	\$375,000.00
			Self	Grant / Contract	Mayo Clinic	Career Development Award supporting 0.1FTE and some research expenses	\$150,000.00
			Self	Employment	Mayo Clinic	Division of Rheumatology, full time, hybrid research and clinical position	
Patil A. Injean	Lit Review Team	Cedars-Sinai				Nothing to disclose	
Nikhil Jiwrajka	Lit Review Team	Penn Medicine	Self	Grant / Contract	Perelman School of Medicine, University of Pennsylvania		\$15,000.00
			Self	Employment	Perelman School of Medicine, University of Pennsylvania		
			Self	Grant / Contract	Perelman School of Medicine, University of Pennsylvania		\$40,000.00
Elena Joerns	Lit Review Team	UT Southwestern Medical Center	Self	Employment	University of Texas Southwestern Medical Center		
			Self	Grant / Contract	Pfizer Inc.	22.7% effort	\$50,000.00

American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease

Project Plan – August 2022

			T32 training grant supporting Elena Joerns' salary	Grant / Contract	National Institute of Health	The major goals of this project are to perform a detailed phenotypic analysis of immunosuppressed patients with interstitial pneumonia with autoimmune features in the UTSW cohort and assess predictors of response to immunosuppression.	\$259,496.00
Joyce Lee	Lit Review Team	University of Colorado	Self	Independent Contractor - Consultant	Boehringer Ingelheim		\$1,200.00
			Self	Grant / Contract	Boehringer Ingelheim	Only start up costs have been received, the milestone based payment of the remainder of the grant has been delayed due to the pandemic.	\$500,000.00
			Self	Grant / Contract	Boehringer Ingelheim	Aryeh Fischer was site PI originally, transferred to me beginning August 2019 after his departure	\$128,954.00
			Self	Grant / Contract	Bristol-Myers Squibb		\$18,320.00
			Self	Grant / Contract	Boehringer Ingelheim	my role is co-investigator	\$491,453.00
			Self	Grant / Contract	Novartis		\$71,253.00
			Self	Independent Contractor - Consultant	Pulmonary Fibrosis Foundation	They support 25% of my time	

American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease

Project Plan – August 2022

			Self	Grant / Contract	FibroGen		\$30,455.00
			Self	Grant / Contract	U.S. Department of Defense	my role is co-investigator	\$9,969,396.00
			Self	Employment	Pulmonary Fibrosis Foundation	They support 25% of my time	
			Self	Grant / Contract	National Heart, Lung, and Blood Institute		\$576,298.00
			Self	Independent Contractor - Consultant	ElevenP15		\$6,000.00
			Self	Independent Contractor - Data And Safety Monitoring	United Therapeutics Corporation		\$1,000.00
			Self	Independent Contractor - Data And Safety Monitoring	SyneosHealth		\$2,000.00
			Self	Independent Contractor - Member of the ILD editorial board	Chest Journal		
			Self	Employment	university of Colorado school of medicine		
			Self	Grant / Contract	UCLA Health System	Aryeh Fischer was site PI originally, transferred to me beginning August 2019 after his departure	\$93,553.00
			Self	Grant / Contract	National Heart, Lung, and Blood Institute	My role is co-investigator	\$6,195,411.00
			Self	Independent Contractor - Co-chair of program committee	American Thoracic Society		
			Self	Grant / Contract	Galapagos	close out ongoing	\$33,291.00
			Self	Independent Contractor - Consultant	United Therapeutics Corporation		\$800.00

American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease

Project Plan – August 2022

Ashima Makol	Lit Review Team	Mayo Clinic	Self	Independent Contractor - Co-investigator for clinical trial sub study	Boehringer Ingelheim	No compensation. Nailfold capillaroscopy substudy complete more than 2 years ago. SENSICIS-ON participants are seen periodically for physical exams.	
			Self	Employment	Mayo Clinic		
			Self	Independent Contractor - Site PI for the KD025-209 study	Kadmon Corporation LLC	No personal compensation. Costs are study execution costs on site. Trial recently closed to recruitment by Sanofi. No further recruitment activity at our site.	\$22,630.00
			Self	Independent Contractor - Medical content contributor	Figure1		\$1,300.00
			Self	Independent Contractor - HZN-825 in Patients With Diffuse Cutaneous Systemic Sclerosis	Horizon Therapeutics plc	Phase Study is not active yet but plan to activate april/may 2022 at our site.	
Gregory McDermott	Lit Review Team	Massachusetts General Hospital	Self	Employment	Partners Healthcare		
			Self	Employment	Brigham and Women's Hospital	Clinical Fellow	
Jake G Natalini	Lit Review Team	NYU Langone Health				Nothing to disclose	
Justin Oldham	Lit Review Team	University of California, Davis	Self	Independent Contractor - Consultant	Boehringer Ingelheim		\$20,000.00
			Self	Independent Contractor - Consultant	United Therapeutics Corporation		\$3,000.00
			Self	Independent Contractor - Consultant	F. Hoffmann-La Roche		\$5,800.00
			Self	Independent Contractor - Consultant	Lupin Pharmaceuticals, Inc		\$5,000.00
Didem Saygin	Lit Review Team	University of Chicago				Nothing to disclose	



American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease

Project Plan – August 2022

Kimberly Showalter Lakin	Lit Review Team	Hospital for Special Surgery, Weill Cornell Medicine				Nothing to disclose	
Namrata Singh	Lit Review Team	University of Washington	Self	Grant / Contract	American Heart Association		\$200,000.00
			Self	Grant / Contract	Rheumatology Research Foundation		\$375,000.00
Joshua J. Solomon	Lit Review Team	National Jewish Health	Self	Independent Contractor - Investigator Initiated Grant Funding	Boehringer Ingelheim	30,000 total grant funding for 2022	\$30,000.00
			Self	Independent Contractor - CTD ILD education	Boehringer Ingelheim		\$6,000.00
			Self	Independent Contractor - PI TRAIL1 Trial	Genentech		\$14,000.00
			Self	Grant / Contract	PFIZER CANADA INC		\$82,904.00
Jeffrey Sparks	Lit Review Team	Brigham and Women's Hospital	Self	Independent Contractor - Consultant	Amgen		
			Self	Independent Contractor - Consultant	Bristol-Myers Squibb		
			Self	Independent Contractor - Consultant	AbbVie		
Marat Turgunbaev	Lit Review Team	American College of Rheumatology				Nothing to disclose	
Samera Vaseer	Lit Review Team	University of Oklahoma				Nothing to disclose	
Rohit Aggarwal	Voting Panel	University of Pittsburgh Medical Center	Self	Independent Contractor - Consultant	Scipher		
			Self	Intellectual Property - Other Intellectual Property		Under University of Pittsburgh	
			Self	Independent Contractor - Consultant	Kezar		
			Self	Independent Contractor - Consultant	Alexion Pharmaceuticals, Inc.		
			Self	Independent Contractor - Consultant	E.R. Squibb & Sons, L.L.C.		
			Self	Grant / Contract	E.R. Squibb & Sons, L.L.C.		\$148,531.84
			Self	Independent Contractor - Consultant	Galapagos		
			Self	Independent Contractor - Consultant	Mallinckrodt LLC		
			Self	Independent Contractor - Consultant	argenx		
			Self	Independent Contractor - Consultant	EMD Serono		
			Self	Independent Contractor - Consultant	corbus		
			Self	Independent Contractor - Consultant	kyverna		
			Self	Independent Contractor - Consultant	Horizon Therapeutics plc		



American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease

Project Plan – August 2022

			Self	Independent Contractor - Consultant	Merck		
			Self	Grant / Contract	Mallinckrodt Hospital Products Inc.		
			Self	Grant / Contract	Q32		\$10,000.00
			Self	Independent Contractor - Consultant	AstraZeneca Pharmaceuticals LP		
			Self	Independent Contractor - Consultant	Roivant		
			Self	Grant / Contract	Mallinckrodt LLC		\$75,000.00
			Self	Grant / Contract	EMD Serono		\$10,000.00
			Self	Independent Contractor - Consultant	Q32		
			Self	Independent Contractor - Consultant	Janssen Global Services, LLC		
			Self	Independent Contractor - Consultant	CSL Behring		
			Self	Independent Contractor - Consultant	Janssen Biotech, Inc.		
			Self	Independent Contractor - Consultant	AbbVie Inc.		
			Self	Independent Contractor - Consultant	Teva Pharmaceutical Industries		
			Self	Independent Contractor - Consultant	Boehringer Ingelheim Pharmaceuticals, Inc.		
			Self	Independent Contractor - Consultant	Octapharma USA, Inc.		
			Self	Independent Contractor - Consultant	Pfizer		
Shervin Assasi	Voting Panel	Texas Health Science Center Houston	Self	Grant / Contract	National Institute of Health		\$783,582.00
			Self	Independent Contractor - Section Editor	Current Opinions in Rheumatology		
			Self	Grant / Contract	Scleroderma Research Foundation		\$467,797.00
			Self	Independent Contractor - Editorial Board	Arthritis Research and Therapy		
			Self	Grant / Contract	National Institute of Health		\$1,710,960.00
			Self	Independent Contractor - Speaker bureau	Integrity continuous education		\$2,500.00
			Self	Employment	The University of Texas Health Science Center at Houston		
			Self	Grant / Contract	National Institute of Health		\$346,497.00
			Self	Independent Contractor - Consultant	AstraZeneca		\$4,296.00
			Self	Independent Contractor - Speaker	North Carolina Rheumatology Association		\$4,000.00



American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease

Project Plan – August 2022

			Self	Independent Contractor - President	scleroderma clinical trial consortium	This is a non-profit, global organization for improving quality of clinical research in systemic sclerosis.	
			Self	Independent Contractor - Consultant	Boehringer Ingelheim	I have not yet received any compensation yet but the estimated compensation for 2022 is \$15,000 to \$20,000.	
			Self	Grant / Contract	Boehringer Ingelheim		\$127,084.00
			Self	Grant / Contract	Boehringer Ingelheim		\$108,383.00
			Self	Independent Contractor - Consultant	Boehringer Ingelheim		\$21,465.59
			Self	Independent Contractor - Consultant	CSL Behring		\$2,115.00
			Self	Grant / Contract	Janssen Biotech, Inc.		\$246,254.00
			Self	Independent Contractor - Consultant	AbbVie		\$2,250.00
Lenore Buckley	Voting Panel	Yale School of Medicine				Nothing to disclose	
Paul F. Dellaripa	Voting Panel	Brigham and Women's Hospital	Self	Independent Contractor - committee member	Food and Drug Administration		
			Self	Independent Contractor - Clinical investigator	Genentech		
			Self	Independent Contractor - Clinical investigator	Bristol-Myers Squibb	no payment of money to me	
			Self	Independent Contractor - committee member	Boehringer Ingelheim		
Robyn T. Domsic	Voting Panel	University of Pittsburgh Medical Center	Self	Independent Contractor - Consultant	CSL Behring		
Tracy Doyle	Voting Panel	Brigham and Women's Hospital	Self	Grant / Contract	Genentech	Site PI for TRAIL1 clinical trial (0.12 calendar months or 1% effort). \$9,750 total cost, \$7,500 direct cost per year.	\$9,750.00

American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease

Project Plan – August 2022

Tracy M. Frech	Voting Panel	Vanderbilt University Medical Center				Nothing to disclose	
Monique E. Hinchcliff	Voting Panel	Yale School of Medicine	Self	Grant / Contract	Boehringer Ingelheim		\$950,231.00
Cheilonda Johnson	Voting Panel	Penn Medicine				Nothing to disclose	
Jeffrey P. Kanne	Voting Panel	University of Wisconsin School of Medicine and Public Health	Self	Independent Contractor - Consultant	Calyx.ai	Independent reviewer for clinical trials - 1 hour per week	\$15,000.00
			Self	Independent Contractor - Consultant	Bayer Healthcare	Intermittent clinical trial work	\$5,000.00
			Self	Independent Contractor - Consultant	Elsevier		\$1,000.00
			Self	Independent Contractor - Consultant	Delfi Diagnostics	\$350 per hour up to 20 hours	\$475.00
			Self	Independent Contractor - Consultant	Wolters Kluwer Health, Inc.		\$3,000.00
John S. Kim	Voting Panel	University of Virginia School of Medicine	Self	Grant / Contract	Pulmonary Fibrosis Foundation		\$50,000.00
			Self	Grant / Contract	National Heart, Lung, and Blood Institute		\$171,612.00
Scott Matson	Voting Panel	University of Kansas Medical Center	Self	Employment	School of Medicine, University of Kansas		
			Self	Independent Contractor - Consultant	imvaria diagnostics		
Zsuzsanna McMahan	Voting Panel	Johns Hopkins University School of Medicine	Self	Grant / Contract	Jerome L. Greene Foundation		\$50,000.00
			Self	Grant / Contract	National Institute of Health		\$149,703.00
			Self	Grant / Contract	Corbus Pharmaceuticals Holdings	I was a sub-PI who did skin biopsies; I did get salary support for each biopsy I perform (~1% total)	\$1,200.00
			Self	Independent Contractor - ACR Representative to the Ex Officio Advisory Board	World Scleroderma Foundation		
			Self	Intellectual Property - Other Intellectual Property			



American College of Rheumatology (ACR) Guideline for the Screening, Monitoring, and Treatment of Interstitial Lung Disease in People with Systemic Autoimmune Rheumatic Disease

Project Plan – August 2022

Lee Shapiro	Voting Panel	Albany Medical College				Nothing to disclose	
Christine D. Sharkey	Voting Panel	University of Wisconsin School of Medicine and Public Health				Nothing to disclose	
Ross S. Summer	Voting Panel	Thomas Jefferson University Hospital	Self	Independent Contractor - Horizon Trial IPF	Horizon Pharma plc		
John Varga	Voting Panel	University of Michigan	Self	Independent Contractor - Consultant	Emerald pharma		
			Self	Independent Contractor - Consultant	up to date		
			Self	Independent Contractor - editor	current rheumatology reports		
			Self	Independent Contractor - Consultant	Boehringer Ingelheim		\$4,000.00
			Self	Independent Contractor - Consultant	Best doctors		
			Self	Independent Contractor - EIC	WILEY		\$5,000.00
			Self	Independent Contractor - Consultant	Boehringer Ingelheim		
			Self	Independent Contractor - Consultant	TeneoBio Pharma		\$20,000.00