Online Supplement:

**Manuscript Title:**
Prognosis of adults with idiopathic pulmonary fibrosis without anti-fibrotic therapy: a systematic review

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S1. Database search strategy

**MEDLINE (Ovid)**

1. Idiopathic Pulmonary Fibrosis/
2. ((pulmonary$ or lung$ or alveoli$) adj2 (fibros$ or fibrot$)).tw.
3. 1 or 2
4. Randomized Controlled Trial.pt.
5. (randomized or randomised).ab,ti.
6. placebo.ab,ti.
7. dt.fs.
8. randomly.ab,ti.
9. trial.ab,ti.
10. groups.ab,ti.
11. or/4-10
12. follow-up studies.sh.
13. cohort.tw.
14. exp mortality/
15. course$.tw.
16. prognos$.tw.
17. predict$.tw.
18. incidence.sh.
19. survival analysis/
20. or/12-19
21. 3 and (11 or 20)
22. Animals/
23. Humans/
24. 22 not (22 and 23)
25. 21 not 24

**CINAHL (EBSCO)**

S1 (MH "Idiopathic Pulmonary Fibrosis")
S2 idiopathic* AND (pulmonary* OR lung* OR alveoli*) AND (fibros* OR fibrot*)
S3 S1 OR S2
S4 (DE "RANDOMIZED CONTROLLED TRIALS")
S5 randomized or randomised
S6 randomly
S7 placebo*
S8 groups
S9 S4 OR S5 OR S6 OR S7 OR S8
S10 (MH "Prospective Studies+)")
S11 cohort
S12 (MH "Mortality+)")
S13 course*
S14 prognos*
S15 predict*
S16 (MH "incidence")
S17 (MH "Survival Analysis+")
S18 S10 OR S11 OR S12 OR S13 OR S14 OR S15 OR S16 OR S17
S19 S3 AND (S9 OR S18)
Embase (Ovid)
1. fibrosing alveolitis/
2. (idiopathic$ adj2 (pulmonary$ or lung$ or alveoli$) adj2 (fibros$ or fibrot$)).tw.
3. 1 or 2
4. Randomized Controlled Trial/
5. randomization/
6. (clinica$ adj3 trial$).tw.
7. exp Placebo/
8. placebo$.ti,ab.
9. random$.ti,ab.
10. groups.ti,ab.
11. or/4-10
12. follow up/
13. cohort.tw.
14. exp mortality rate/
15. course$.tw.
16. prognos$.tw.
17. predict$.tw.
18. incidence/
19. survival analysis/
20. or/12-19
21. 3 and (11 or 20)
22. exp animals/ or exp invertebrate/ or animal experiment/ or animal model/ or animal tissue/ or animal cell/ or nonhuman/
23. human/ or normal human/ or human cell/
24. 22 not (22 and 23)
25. 21 not 24

CENTRAL (Cochrane Register of Studies Online)
#1 MESH DESCRIPTOR Idiopathic Pulmonary Fibrosis EXPLODE ALL TREES
#2 ((pulmonary* or lung* or alveoli*) adj2 (fibros* or fibrot*)):TI,AB,KY
#3 #1 OR #2

PubMed
("idiopathic pulmonary fibrosis"[MeSH Terms] OR "pulmonary fibrosis"[Title/Abstract]) AND
((randomized controlled trial[Publication Type] OR (randomized[Title/Abstract] AND controlled[Title/Abstract] AND trial[Title/Abstract])) OR (incidence[MeSH:noexp] OR mortality[MeSH Terms] OR follow up studies[MeSH:noexp] OR prognos*[Text Word] OR predict*[Text Word] OR course*[Text Word]))
### S2. Risk of bias assessment tool

| Criteria                                      | High risk of bias | Low risk of bias | Unclear/Other |
|-----------------------------------------------|-------------------|------------------|---------------|
| Sample                                        | Clinical          | Population       | Unclear       |
| Recruitment                                   | Retrospective     | Prospective      | Unclear       |
| Selection criteria for participants<sup>a</sup> | No                | Yes              | Unclear       |
| Baseline characteristics of participants<sup>*</sup> | No                | Yes              | Unclear       |
| Follow-up percentage (if RCT)                 | < 80%             | ≥ 80%            | Unclear       |
| Follow-up duration                            | < 1 year          | ≥ 1 year         | Unclear       |
| Reason lost to follow-up                     | No                | Yes              | Unclear       |
| Timing of diagnosis                           | At the conclusion of the study | At baseline or before recruitment to study | Unclear |
| Blinding (if RCT)                             | Not blinded       | Blinding adequate | Unclear       |
| Outcome described a priori                    | No                | Yes              | Unclear       |
| Intention-to-treat analysis (if RCT)           | No                | Yes              | Unclear       |
| Adequate description of statistical analysis  | No                | Yes              | Unclear       |

<sup>a</sup> Scoring was based on the description of the inclusion and exclusion criteria of participants: low risk if adequately described; unclear if inadequately described; high risk if not described

<sup>*</sup> Scoring was based on the description of baseline characteristics of participants: low risk if adequately described; unclear if inadequately described; high risk if not described
### S3. Reasons of exclusion and references for excluded studies

#### Part A: Reason for exclusion

| Study                  | Reason for exclusion                                                                 |
|------------------------|---------------------------------------------------------------------------------------|
| Abe 2012               | Follow-up duration < 12 months                                                        |
| Abe 2015               | Follow-up duration < 12 months                                                        |
| Abhyankar 2013         | Follow-up duration unclear; author can’t be contacted                                  |
| Abu-Hussein 2010       | Follow-up duration unclear; author can’t be contacted                                  |
| Acar Silva 2013        | Follow-up duration unclear; author can’t be contacted                                  |
| Acosta Fernández 2006  | Editorial                                                                             |
| Adamali 2012           | Follow-up duration unclear; author can’t be contacted                                  |
| Aggarwal 2016          | Not a randomised controlled trial or cohort study                                      |
| Aggarwal 2017          | Follow-up duration unclear; author can’t be contacted                                  |
| Agostini 2005          | Review article                                                                        |
| Alghamdi 2015          | Follow-up duration unclear; author can’t be contacted                                  |
| Alhamad 2015a          | Follow-up duration unclear; author can’t be contacted                                  |
| Alhamad 2015           | Follow-up duration unclear; author can’t be contacted                                  |
| Alhamad 2012           | Follow-up duration unclear; author can’t be contacted                                  |
| Alhamad 2012a          | Follow-up duration unclear; author can’t be contacted                                  |
| Alhamad 2015           | Included patients receiving antifibrotic agents (nintedanib or pirfenidone)           |
| Alhamad 2015a          | Follow-up duration unclear; author can’t be contacted                                  |
| Alhamad 2016           | Follow-up duration unclear; author can’t be contacted                                  |
| Alton 1989              | Participants overlap with other included study (Agusti 1994)                           |
| Ambur 2015              | Follow-up duration unclear; author can’t be contacted                                  |
| Alken 2015             | Not idiopathic pulmonary fibrosis                                                      |
| Andersen 2012          | Follow-up duration unclear; author can’t be contacted                                  |
| Ando 2011              | Follow-up duration unclear; author can’t be contacted                                  |
| Ando 2011a             | Foreign language (translation not available)                                          |
| Ando 2013              | Follow-up duration variable; author can’t be contacted                                 |
| Anonymous 1971         | Not idiopathic pulmonary fibrosis                                                      |
| Anonymous 1978         | Editorial                                                                             |
| Anonymous 1995         | Foreign language (translation not available)                                          |
| Anonymous 1999         | Review article                                                                        |
| Anonymous 2005         | Patient education handout                                                              |
| Anonymous 2006         | Review article                                                                        |
| Anonymous 2017         | Not a randomised controlled trial or cohort study                                      |
| Ansarie 2012           | Mixed disease group                                                                   |
| Antoniou 2002          | No placebo group                                                                      |
| Antoniou 2003          | No placebo group                                                                      |
| Antoniou 2004          | No placebo group                                                                      |
| Antoniou 2006          | No placebo group                                                                      |
| Reference                        | Detailed Description                                                                 |
|---------------------------------|--------------------------------------------------------------------------------------|
| Antoniou 2012                   | Follow-up duration unclear; author can’t be contacted                                 |
| Appel 2007                      | Follow-up duration unclear; author can’t be contacted                                 |
| Arai 2012                       | Included patients receiving antifibrotic agents (nintedanib or pirfenidone)           |
| Arai 2013                       | Follow-up duration unclear; author can’t be contacted                                 |
| Arai 2014                       | Included patients receiving antifibrotic agents (nintedanib or pirfenidone)           |
| Arai 2015                       | Follow-up duration unclear; author can’t be contacted                                 |
| Arai 2016                       | Follow-up duration unclear; author can’t be contacted                                 |
| Arai 2016a                      | Follow-up duration unclear; author can’t be contacted                                 |
| Arango Tomas 2015               | Follow-up duration < 12 months                                                       |
| Arizono 2015                    | Not a randomised controlled trial or cohort study                                     |
| Ash 2017                        | Follow-up duration unclear; author can’t be contacted                                 |
| Assayag 2014                    | Mixed disease group                                                                  |
| Assayag 2015                    | Mixed disease group                                                                  |
| Aubry 2002                      | Follow-up duration unclear; author can’t be contacted                                 |
| Awano 2017                      | Not a randomised controlled trial or cohort study                                     |
| Baba 2010                       | Follow-up duration unclear; author can’t be contacted                                 |
| Baba 2012                       | Follow-up duration unclear; author can’t be contacted                                 |
| Baba 2013                       | Follow-up duration unclear; author can’t be contacted                                 |
| Baddini-Martinez 1993           | Follow-up duration variable; author can’t be contacted                                 |
| Balade Martinez 2014            | Included patients receiving antifibrotic agents (nintedanib or pirfenidone)           |
| Balestro 2012                   | Follow-up duration unclear; author can’t be contacted                                 |
| Balestro 2013                   | Follow-up duration variable; author can’t be contacted                                 |
| Balestro 2014                   | Follow-up duration unclear; author can’t be contacted                                 |
| Balestro 2016                   | Follow-up duration variable; author can’t be contacted                                 |
| Balestro 2016a                  | Included patients receiving antifibrotic agents (nintedanib or pirfenidone)           |
| Bando 2001                      | Follow-up duration variable; author can’t be contacted                                 |
| Bando 2010                      | Possible use of antifibrotic agents (nintedanib or pirfenidone); author can’t be contacted |
| Bando 2012                      | Foreign language - translation not available                                          |
| Bando 2014                      | Included patients receiving antifibrotic agents (nintedanib or pirfenidone)           |
| Bando 2014a                     | Included patients receiving antifibrotic agents (nintedanib or pirfenidone)           |
| Bando 2015a                     | Included patients receiving antifibrotic agents (nintedanib or pirfenidone)           |
| Bando 2016                      | Included patients receiving antifibrotic agents (nintedanib or pirfenidone)           |
| Barber 2014                     | Not a randomised controlled trial or cohort study                                     |
| Barber 2015                     | Not a randomised controlled trial or cohort study                                     |
| Barber 2016                     | Not a randomised controlled trial or cohort study                                     |
| Barbero 2013                    | Included patients receiving antifibrotic agents (nintedanib or pirfenidone)           |
| Barlo 2009a                     | Follow-up duration variable; author can’t be contacted                                 |
| Barry 2011                      | Included patients receiving antifibrotic agents (nintedanib or pirfenidone)           |
| Barry 2012                      | Included patients receiving antifibrotic agents (nintedanib or pirfenidone)           |
| Battista 2003                   | Follow-up duration variable; author can’t be contacted                                 |
| Baughman 1992                   | Follow-up duration variable; author can’t be contacted                                 |
| Behera 1998                     | Follow-up duration variable; author can’t be contacted                                 |
| Behr 2002                       | Mixed disease group                                                                  |
| Behr 2006                       | Review article                                                                      |
| Behr 2014                       | Included patients receiving antifibrotic agents (nintedanib or pirfenidone)           |
| Behr 2014a                      | Not a randomised controlled trial or cohort study                                     |
| Behr 2014b                      | Not a randomised controlled trial or cohort study                                     |
| Behr 2015                       | Included patients receiving antifibrotic agents (nintedanib or pirfenidone)           |
| Behr 2015a                      | Included patients receiving antifibrotic agents (nintedanib or pirfenidone)           |
| Reference                  | Comment                                                                 |
|---------------------------|-------------------------------------------------------------------------|
| Behr 2015b                | Not a randomised controlled trial of cohort study                       |
| Behr 2015c                | Follow-up duration variable; author can’t be contacted                   |
| Behr 2015d                | Follow-up duration variable; author can’t be contacted                   |
| Behr 2016                 | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Beltramo 2016             | Follow-up duration unclear; author can’t be contacted                    |
| Bennett 2017              | Not idiopathic pulmonary fibrosis                                        |
| Benson 1972               | Not idiopathic pulmonary fibrosis                                        |
| Berry 2012                | Mixed disease group                                                     |
| Best 2008                 | Follow-up duration variable; author can’t be contacted                   |
| Black 2013                | Follow-up duration unclear; author can’t be contacted                    |
| Blackburn 2015            | Follow-up duration unclear; author can’t be contacted                    |
| Bodlet 2012               | Follow-up duration variable; author can’t be contacted                   |
| Bodlet 2013               | Follow-up duration variable; author can’t be contacted                   |
| Bois 2016                 | Not a randomised controlled trial or cohort study                       |
| Bollinelli 1973           | Not a randomised controlled trial or cohort study                       |
| Bonella 2013              | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Bonella 2014              | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Bonella 2015              | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Bonella 2015a             | Review article                                                          |
| Bonella 2016              | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Bonella 2016a             | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Bonham 2014               | Follow-up duration unclear; author can’t be contacted                    |
| Boon 2009                 | Not a randomised controlled trial or cohort study                       |
| Borges 2016               | No relevant outcomes                                                    |
| Borie 2011                | Follow-up duration variable; author can’t be contacted                   |
| Borie 2012                | Follow-up duration variable; author can’t be contacted                   |
| Borie 2013                | Follow-up duration variable; author can’t be contacted                   |
| Borie 2016                | Mixed disease group                                                     |
| Bournazos 2009            | Participants overlap with other included study (Bournazos 2011)          |
| Bournazos 2010            | Participants overlap with other included study (Bournazos 2011)          |
| Bournazos 2010a           | Participants overlap with other included study (Bournazos 2011)          |
| Bradford 2003             | Follow-up duration variable; author can’t be contacted                   |
| Bradford 2004             | Follow-up duration variable; author can’t be contacted                   |
| Bradshaw 2012             | Diagnosis unclear; author can’t be contacted                             |
| Britton 2000              | Review article                                                          |
| Broder 2016               | Not a randomised controlled trial or cohort study                       |
| Brown 1971                | Not a randomised controlled trial or cohort study                       |
| Brown 2012                | Participants overlap with other included study (Fisher 2017)             |
| Brown 2015                | Participants overlap with other included study (Fisher 2017)             |
| Brunetti 2013             | Follow-up duration unclear; author can’t be contacted                    |
| Brunnenemer 2016          | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Burgess 2015              | Not a randomised controlled trial or cohort study                       |
| Burgess 2015a             | Not a randomised controlled trial or cohort study                       |
| Burrell 2012              | Follow-up duration unclear; author can’t be contacted                    |
| Callahan 2016             | Participants overlap with other included study (Toelle 2014)             |
| Caminati 2009             | Follow-up duration variable; author can’t be contacted                   |
| Campainha 2011            | Follow-up duration unclear; author can’t be contacted                    |
| Cao 2016                  | Follow-up duration variable; author contacted                           |
| Capano 2015               | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Carbone 2006              | Follow-up duration variable; author can’t be contacted                   |
| Carbone 2010              | Follow-up duration unclear; author can’t be contacted                    |
| Carbone 2010a             | Follow-up duration unclear; author can’t be contacted                    |
| Reference          | Description                                                                 |
|--------------------|------------------------------------------------------------------------------|
| Caro 2016          | Follow-up duration variable; author can’t be contacted                        |
| Casanova 2009      | Foreign language (translation not available)                                  |
| Cegla 1974         | Follow-up duration < 12 months                                               |
| Cegla 1976         | Paper not available for review                                               |
| Cegla 1977         | Not a randomised controlled trial or cohort study                             |
| Cegla 1980         | Not a randomised controlled trial or cohort study                             |
| Cerri 2012         | Follow-up duration < 12 months                                               |
| Cerri 2013         | Follow-up duration unclear; author can’t be contacted                         |
| Chan 1997          | Participants overlap with other included study (Jacob 2016)                  |
| Chartrand 2015     | Participants overlap with other included study (Strand 2014)                 |
| Chaudhuri 2014     | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Chaudhuri 2014a    | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Chen 2012          | Mixed disease group                                                           |
| Chetta 2005        | Editorial                                                                     |
| Chien 2012         | Follow-up duration unclear; author can’t be contacted                         |
| Chien 2013         | Follow-up duration variable; author can’t be contacted                         |
| Chien 2014         | Follow-up duration variable; author can’t be contacted                         |
| Chodosowska 1988   | Included participants < 18 years old                                         |
| Choi 2014          | Participants overlap with other included study (Richards 2012)               |
| Chuchalin 2000     | Review article                                                                |
| Churg 2007         | Mixed disease group                                                           |
| Cinel 2010         | Not adult population                                                          |
| Cioffi Squitieri 2014 | Mixed disease group                                                      |
| Claar 2015         | Not a randomised controlled trial or cohort study                             |
| Cohen 2013         | Follow-up duration < 12 months                                               |
| Coll 2011          | Foreign language (translation not available)                                  |
| Collard 2003       | Participants overlap with other included study (Strand 2014)                 |
| Collard 2004       | Participants overlap with other included study (Strand 2014)                 |
| Collard 2007       | Participants overlap with other included study (Strand 2014)                 |
| Collard 2010       | Editorial                                                                     |
| Collard 2012a      | Follow-up duration variable                                                   |
| Collard 2014       | Participants overlap with other included study (STEP-IPF/ACE-IPF/PANTHER-IPF trials) |
| Colombi 2015       | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Colombi 2015a      | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Colombi 2016       | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Condos 2016        | Not a randomised controlled trial or cohort study                             |
| Corte 2009         | Mixed disease group                                                           |
| Corte 2010         | Participants overlap with other included study (Jacob 2016)                  |
| Corte 2012         | Participants overlap with other included study (Jacob 2016)                  |
| Corte 2012a        | Mixed disease group                                                           |
| Corte 2013         | Comment                                                                       |
| Corte 2014         | Participants overlap with other included study (Jo 2017)                     |
| Corte 2014a        | Participants overlap with other included study (Jo 2017)                     |
| Corte 2015         | Not a randomised controlled trial or cohort study                             |
| Corte 2016         | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Costa da Silva 2009 | Mixed disease group                                                          |
| Costabel 2011      | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Costabel 2012      | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Costabel 2014      | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Costabel 2016      | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Costabel 2016a     | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Reference                | Overview                                                                 |
|--------------------------|---------------------------------------------------------------------------|
| Costabel 2016b           | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| CoTherix 2005            | Follow-up duration < 12 months                                            |
| Cottin 2012              | Mixed disease group                                                       |
| Cottin 2013              | Mixed disease group                                                       |
| Cottin 2013a             | Mixed disease group                                                       |
| Cottin 2015              | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Cottin 2015a             | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Coultas 1996a            | Paper not available for review                                            |
| Craig 2014               | Follow-up duration unclear; author can’t be contacted                      |
| Crestani 2012            | Follow-up duration < 12 months                                            |
| Crestani 2015            | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Crestani 2015a           | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Crestani 2015b           | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Crestani 2015c           | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Crestani 2016            | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| D’Andrea 2016            | Uncertain exclusion criteria for use of antifibrotic agents; author can’t be contacted |
| Dabar 2014               | Follow-up duration unclear; author can’t be contacted                      |
| Dai 2015                 | Follow-up duration unclear; author can’t be contacted                      |
| Dai 2015a                | Not a randomised controlled trial or cohort study                         |
| Dalleywater 2014         | Follow-up duration variable; author can’t be contacted                     |
| Dalleywater 2015         | Follow-up duration variable; author can’t be contacted                     |
| Daniil 1999              | Follow-up duration variable; author can’t be contacted                     |
| Dawson 2012              | Review article                                                            |
| Dayton 1993              | Follow-up duration variable; author can’t be contacted                     |
| de Andrade 2010          | Follow-up duration variable; author can’t be contacted                     |
| de Cremoux 1990          | Follow-up duration unclear; author can’t be contacted                      |
| de Lauretis 2010         | Participants overlap with other included study (Jacob 2016)               |
| de Lauretis 2013         | Participants overlap with other included study (Jacob 2016)               |
| De Meester 1999          | Mixed disease group                                                       |
| De Meester 2001          | Mixed disease group                                                       |
| de W Kitcat 1928         | Not idiopathic pulmonary fibrosis                                          |
| de Wall 1991             | Diagnosis unclear; author can’t be contacted                               |
| DePianto 2015            | Follow-up duration unclear                                                |
| DePianto 2016            | Follow-up duration unclear                                                |
| Devaraj 2009             | Follow-up duration unclear                                                |
| Dierkesmann 1972         | Patient age unclear; author can’t be contacted                             |
| Dietzsch 1966            | Not a randomised controlled trial or cohort study                          |
| Dimadi 2003              | No placebo group                                                          |
| Dimmock 2013             | Follow-up duration unclear; author can’t be contacted                      |
| Dimmock 2013a            | Not a randomised controlled trial or cohort study                          |
| Divihotawela 2016        | Follow-up duration unclear                                                |
| Doherty 1997             | Follow-up duration unclear; author can’t be contacted                      |
| Donahoe 2015             | Follow-up duration variable                                               |
| Dong 2015                | Follow-up duration unclear; author can’t be contacted                      |
| Doubkova 2014            | Follow-up duration unclear; author can’t be contacted                      |
| Doubkova 2016            | Follow-up duration unclear; author can’t be contacted                      |
| Doubkova 2016a           | Follow-up duration unclear; author can’t be contacted                      |
| Douglas 1997             | Follow-up duration unclear; author can’t be contacted                      |
| Douglas 1998             | No placebo group                                                          |
| Douglas 2000             | Follow-up duration unclear; author can’t be contacted                      |
| Downman 2016             | Not a randomised controlled trial or cohort study                          |
| Study                | Description                                                                 |
|---------------------|-----------------------------------------------------------------------------|
| du Bois 2011        | Follow-up duration < 12 months                                              |
| du Bois 2013        | Follow-up duration unclear; author can’t be contacted                       |
| du Bois 2014        | Letter                                                                       |
| Duong 2015          | Follow-up duration unclear; author can’t be contacted                       |
| Durheim 2015        | Follow-up duration variable                                                 |
| Durheim 2015a       | Follow-up duration variable                                                 |
| Dwarkanath 2013     | Follow-up duration unclear; author can’t be contacted                       |
| Eaden 2015          | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Egan 1996           | Review article                                                              |
| Ekstrom 2016        | Mixed disease group; author contacted                                       |
| Eldahdouh 2017      | Not a randomised controlled trial or cohort study                           |
| Elshafi 2012        | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Elshafi 2015        | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Enomoto 2006        | Follow-up duration variable                                                 |
| Enomoto 2012        | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Enomoto 2013        | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Enomoto 2013a       | Participants overlap with other included study (Fujimoto 2012)              |
| Enomoto 2014        | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Enomoto 2014a       | Participants overlap with other included study (Fujimoto 2012)              |
| Enomoto 2015        | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Enomoto 2015a       | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Fakharian 2010      | Follow-up duration unclear; author can’t be contacted                       |
| Fasano 1999         | Foreign language - translation not available                                |
| Fell 2009           | Follow-up duration unclear                                                  |
| Fernandez 2010      | Follow-up duration unclear; author can’t be contacted                       |
| Finn 2015           | No relevant outcomes                                                        |
| Fischer 2006        | Participants overlap with other included study (Strand 2014)                |
| Fischer 2012        | Follow-up duration unclear; author can’t be contacted                       |
| Flaherty 2000       | Follow-up duration < 12 months                                              |
| Flaherty 2001       | Participants overlap with other included study (Gay 1998)                   |
| Flaherty 2004       | Follow-up duration < 12 months                                              |
| Flaherty 2006       | Participants overlap with other included study (Gay 1998)                   |
| Ford-Sahibzada 2016 | Mixed disease group                                                         |
| Franquét 2000       | Follow-up duration < 12 months                                              |
| François 2015       | Follow-up duration unclear; author can’t be contacted                       |
| Fremer 2006         | Follow-up duration unclear; author can’t be contacted                       |
| Fujimoto 2003       | Follow-up duration unclear; author can’t be contacted                       |
| Fujiwara 2006       | Foreign language - translation not available                                |
| Fukihara 2015       | Follow-up duration unclear; author can’t be contacted                       |
| Furukawa 2013       | Follow-up duration unclear; author can’t be contacted                       |
| Furukawa 2015       | Follow-up duration unclear; author can’t be contacted                       |
| Furukawa 2017       | Possible use of antifibrotic agents (nintedanib or pirfenidone); author can’t be contacted |
| Gainza 2017         | Paper not available for review                                              |
| Gandhi 2014         | Follow-up duration variable; author can’t be contacted                       |
| George 2015         | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Gerke 2010          | Mixed disease group                                                         |
| Gerke 2010a         | Mixed disease group                                                         |
| Giddings 2012       | Insufficient data available                                                 |
| Gilani 2010         | Participants overlap with other included study (Richards 2012)              |
| Gille 2016          | Follow-up duration unclear; author can’t be contacted                       |
| Giaspale 2014       | Not a randomised controlled trial or cohort study                           |
| Reference   | Description                                                                                       |
|-------------|--------------------------------------------------------------------------------------------------|
| Glaspole 2015 | Not a randomised controlled trial or cohort study                                                 |
| Glaspole 2016 | Not a randomised controlled trial or cohort study                                                 |
| Glaspole 2016a | Included patients receiving antifibrotic agents (nintedanib or pirfenidone)                    |
| Glaspole 2017 | Included patients receiving antifibrotic agents (nintedanib or pirfenidone)                    |
| Goh 2011     | Participants overlap with other included study (Jacob 2016)                                      |
| Goh 2016     | Not a randomised controlled trial or cohort study                                                 |
| Golec 2008   | Mixed disease group                                                                               |
| Gomes 2015   | Participants overlap with other included study (Soares 2013)                                      |
| Gonzalez 2016 | Follow-up duration unclear; author can’t be contacted                                            |
| Goobie 2016  | Follow-up duration unclear; author can’t be contacted                                             |
| Gottlieb 2010| Mixed disease group                                                                               |
| Gottlieb 2012| Mixed disease group                                                                               |
| Goyard 2015 | Paper not available for review                                                                   |
| Gracey 1970  | Not a randomised controlled trial or cohort study                                                 |
| Greene 2001  | Follow-up duration variable; author can’t be contacted                                            |
| Greene 2002  | Follow-up duration variable; author can’t be contacted                                            |
| Gribbin 2006 | Follow-up duration variable; author contacted                                                     |
| Grijm 2005   | Mixed disease group                                                                               |
| Gruden 2016  | Follow-up duration variable                                                                      |
| Guenther 2014| Not a randomised controlled trial or cohort study                                                 |
| Gurioli 2011 | Follow-up duration variable; author can’t be contacted                                            |
| Guschall 1998| Included participants < 18 years old                                                              |
| Hamai 2016   | Follow-up duration variable                                                                      |
| Hamdy 2014   | Not a randomised controlled trial or cohort study                                                 |
| Hamm 1970    | Review article                                                                                   |
| Han 2002     | Follow-up duration < 12 months                                                                   |
| Han 2008     | Participants overlap with other included study (Gay 1998)                                         |
| Han 2010     | Not a randomised controlled trial or cohort study                                                 |
| Han 2011     | Follow-up duration < 12 months                                                                   |
| Han 2013     | Follow-up duration < 12 months                                                                   |
| Han 2014     | Follow-up duration < 12 months                                                                   |
| Hanak 2008   | Follow-up duration variable                                                                      |
| Hara 2015    | Review article                                                                                   |
| Hara 2016    | Follow-up duration unclear; author can’t be contacted                                             |
| Hara 2017    | Not a randomised controlled trial or cohort study                                                 |
| Harada 2013  | Participants overlap with other included study (Akagi 2009)                                        |
| Harari 1997  | Follow-up duration unclear; author can’t be contacted                                             |
| Harari 2005  | Review article                                                                                   |
| Harari 2014  | Included patients receiving antifibrotic agents (nintedanib or pirfenidone)                      |
| Harari 2015  | Included patients receiving antifibrotic agents (nintedanib or pirfenidone)                      |
| Harari 2015a | Follow-up duration unclear; author can’t be contacted                                             |
| Harari 2017  | Letter                                                                                           |
| Harris 1998  | Not a randomised controlled trial or cohort study                                                 |
| Hashemi 2013 | Follow-up duration variable; author can’t be contacted                                            |
| Hashimoto 2014 | Follow-up duration variable; author can’t be contacted                                           |
| Hayakawa 2015| Included patients receiving antifibrotic agents (nintedanib or pirfenidone)                      |
| Hayakawa 2016| Included patients receiving antifibrotic agents (nintedanib or pirfenidone)                      |
| Hayes 2016   | Follow-up duration variable; author can’t be contacted                                             |
| He 2005      | Follow-up duration < 12 months                                                                   |
| He 2016      | Follow-up duration unclear; author can’t be contacted                                             |
| Herazo 2011  | Follow-up duration unclear                                                                        |
| Study                          | Comments                                                                 |
|-------------------------------|--------------------------------------------------------------------------|
| Herazo-Maya 2013              | Participants overlap with other included study (Richards 2012)           |
| Herazo-Maya 2013a             | Follow-up duration unclear                                               |
| Herazo-Maya 2014              | Participants overlap with other included study (Richards 2012)           |
| Herazo-Maya 2015              | Participants overlap with other included study (Richards 2012)           |
| Herazo-Maya 2015a             | Participants overlap with other included study (Richards 2012)           |
| Herridge 2016                 | Follow-up duration unclear; author can’t be contacted                    |
| Hirano 2017                   | Follow-up duration unclear; author can’t be contacted                    |
| Hiwatari 1991                 | Participants overlap with other included study (Hiwatari 1997)           |
| Hiwatari 1994                 | Not a randomised controlled trial or cohort study                        |
| Ho 2013                       | Participants overlap with other included study (Su 2011)                 |
| Hogaboam 2012                 | Review article                                                          |
| Homma 1995                    | Mixed disease group                                                     |
| Homma 2013                    | Follow-up duration unclear; author can’t be contacted                    |
| Hook 2010                     | Follow-up duration variable; author can’t be contacted                   |
| Hook 2011                     | Follow-up duration variable; author can’t be contacted                   |
| Hook 2012                     | Follow-up duration variable; author contacted                            |
| Hope-Gill 2012                | Follow-up duration unclear; author can’t be contacted                    |
| Horimasu 2011                 | Follow-up duration unclear                                              |
| Horita 2011                   | Follow-up duration < 12 months                                           |
| Hosenpud 1998                 | Mixed disease group                                                     |
| Hosoda 2013                   | Follow-up duration < 12 months                                           |
| Hou 2001                      | No placebo group                                                        |
| Hou 2011                      | Follow-up duration unclear; author can’t be contacted                    |
| Hozumi 2016                   | Follow-up duration variable                                              |
| Huang 2011                    | Follow-up duration unclear                                              |
| Huang 2015                    | Participants overlap with other included study (Oldham 2015)             |
| Huang 2015a                   | Participants overlap with other included study (Oldham 2015)             |
| Hubbard 1996                  | Follow-up duration unclear; author can’t be contacted                    |
| Hubbard 2000                  | Follow-up duration < 12 months                                           |
| Hubbard 2002                  | Follow-up duration unclear; author can’t be contacted                    |
| Huie 2010                     | Mixed disease group                                                     |
| Huie 2011                     | Follow-up duration unclear; author can’t be contacted                    |
| Huppmann 2013                 | Mixed disease group                                                     |
| Hutchinson 2014               | Not a randomised controlled trial or cohort study                        |
| Hwang 2011                    | Mixed disease group                                                     |
| Hyldgaard 2013                | Follow-up duration unclear; author can’t be contacted                    |
| Hyldgaard 2013a               | Follow-up duration unclear; author can’t be contacted                    |
| Hyldgaard 2014                | Follow-up duration unclear; author can’t be contacted                    |
| Hyldgaard 2014a               | Follow-up duration unclear; author can’t be contacted                    |
| Hyldgaard 2015                | Follow-up duration unclear; author can’t be contacted                    |
| Ichimura 2014                 | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Ichimura 2014a                | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Ichimuray, 2014               | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Ihle 2014                     | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Ikeda 2013                    | No relevant outcomes                                                    |
| Ikezoe 2013                   | Follow-up duration unclear; author can’t be contacted                    |
| Ikezoe 2014                   | Follow-up duration variable; author can’t be contacted                   |
| Ishii 2013                    | Follow-up duration variable; author can’t be contacted                   |
| Iwamoto 2014                  | Follow-up duration unclear                                              |
| Iwasawa 2006                  | Foreign language (translation not available)                             |
| Iwasawa 2008                  | Paper not available for review                                          |
| Iwasawa 2009                  | Participants overlap with other included study (Iwasawa 2014)            |
| Reference               | Note                                                                 |
|-------------------------|----------------------------------------------------------------------|
| Iwasawa 2010            | Follow-up duration variable                                         |
| Iwasawa 2017            | Follow-up duration variable                                         |
| Izdebska-Makosa 1981    | Included participants < 18 years old                                |
| Jacob 2016a             | Follow-up duration unclear; author can’t be contacted               |
| Jagadeesan 2016         | No relevant outcomes                                                |
| Jamal 2013              | Follow-up duration unclear; author can’t be contacted               |
| Jankowich 2010          | Mixed disease group                                                 |
| Jegal 2005              | Follow-up duration variable; author can’t be contacted               |
| Jenkins 2015            | Follow-up duration unclear                                          |
| Jeong 2005              | Follow-up duration unclear; author can’t be contacted               |
| Jeze 1984               | Review article                                                      |
| Jeze 1988               | Not idiopathic pulmonary fibrosis                                    |
| Jia 2016                | Follow-up duration unclear; author can’t be contacted               |
| Jiang 2007              | Review article                                                      |
| Jiang 2016              | Mixed disease group                                                 |
| Jindal 1979             | Mixed disease group                                                 |
| Jo 2013                 | Mixed disease group                                                 |
| Jo 2013a                | Mixed disease group                                                 |
| Jo 2015                 | Participants overlap with other included study (Jo 2017)            |
| Jo 2015a                | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Jo 2016                 | Participants overlap with other included study (Jo 2017)            |
| Jo 2016a                | Participants overlap with other included study (Jo 2017)            |
| Jo 2016b                | Participants overlap with other included study (Jo 2017)            |
| Jo 2016c                | Participants overlap with other included study (Jo 2017)            |
| Johnson 1989            | No placebo group                                                    |
| Johnston 1993           | Not a randomised controlled trial or cohort study                   |
| Johnston 1997           | Participants overlap with other included study (Rudd 2007)          |
| Judge 2010              | Follow-up duration unclear; author can’t be contacted               |
| Judge 2012              | Follow-up duration unclear; author can’t be contacted               |
| Kaarteenaho-Wiik 1996   | Follow-up duration variable; author can’t be contacted               |
| Kaddah 2016             | Not a randomised controlled trial or cohort study                   |
| Kadikar 1997            | Follow-up duration unclear; author can’t be contacted               |
| Kagami 2014             | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Kahloon 2013            | Follow-up duration unclear                                          |
| Kakugawa 2016           | Follow-up duration unclear; author can’t be contacted               |
| Kalra 2003              | Follow-up duration unclear                                          |
| Kane 2016               | Not a randomised controlled trial or cohort study                   |
| Kappos 1977             | Not a randomised controlled trial or cohort study                   |
| Kass 2013               | Participants overlap with other included study (Richards 2012)      |
| Kataoka 2014            | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Kato 2016               | Not a randomised controlled trial or cohort study                   |
| Kawabata 2003           | Participants overlap with other included study (Hamada 2007)        |
| Kawakami 1980           | Not a randomised controlled trial or cohort study                   |
| Kawatani 2007           | Foreign language (translation not available)                        |
| Kaya 2015               | Follow-up duration unclear; author can’t be contacted               |
| Keir 2012               | Not idiopathic pulmonary fibrosis                                    |
| Keir 2014               | Not idiopathic pulmonary fibrosis                                    |
| Khadawardi 2016         | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Kim 1994                | Follow-up duration < 12 months                                      |
| Kim 2011                | Mixed disease group                                                 |
| Reference        | Details                                                                 |
|------------------|-------------------------------------------------------------------------|
| Kim 2012a        | Follow-up duration unclear; author can’t be contacted                   |
| Kim 2014a        | Participants overlap with other included study (Song 2011)              |
| Kim 2014b        | Follow-up duration unclear; author can’t be contacted                   |
| Kim 2014c        | Follow-up duration unclear; author can’t be contacted                   |
| Kim 2014d        | Follow-up duration unclear; author can’t be contacted                   |
| Kim 2015a        | Participants overlap with other included study (Song 2011)              |
| Kim 2015b        | Mixed disease group                                                    |
| Kim 2015c        | Participants overlap with other included study (Song 2011)              |
| Kim 2015d        | Not idiopathic pulmonary fibrosis                                       |
| Kim 2016         | No relevant outcomes                                                   |
| Kimura 2013      | Possible use of antifibrotic agents (nintedanib or pirfenidone); author can’t be contacted |
| Kinder 2008      | Participants overlap with other included study (Strand 2014)            |
| Kinder 2009      | Participants overlap with other included study (Strand 2014)            |
| Kinder 2010      | Follow-up duration < 12 months                                         |
| King 2001a       | Participants overlap with other included study (Strand 2014)            |
| King 2001b       | Participants overlap with other included study (Strand 2014)            |
| King 2005        | Not a randomised controlled trial or cohort study                       |
| King 2013        | Not a randomised controlled trial or cohort study                       |
| King 2014b       | Not a randomised controlled trial or cohort study                       |
| King 2014c       | Not a randomised controlled trial or cohort study                       |
| Kinney 2013      | Follow-up duration < 12 months                                         |
| Kinoshita 2013   | Follow-up duration unclear; author can’t be contacted                   |
| Kirk 1984a       | Participants overlap with other included study (Jacob 2016)             |
| Kishaba 2012     | Follow-up duration unclear; author can’t be contacted                   |
| Kishaba 2014     | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Kishaba 2014a    | Follow-up duration unclear; author can’t be contacted                   |
| Kishaba 2015     | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Kishaba 2015a    | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Kishaba 2017     | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Kitamura 2012    | Follow-up duration unclear; author can’t be contacted                   |
| Kitamura 2012a   | Follow-up duration unclear; author can’t be contacted                   |
| Kogan 1995       | Not a randomised controlled trial or cohort study                       |
| Kohashi 2016     | Follow-up duration unclear; author can’t be contacted                   |
| Kohlhaeufl 2014  | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Kokuho 2013      | Review article                                                         |
| Kolb 1998a       | Age of study population unclear                                         |
| Kolb 2016        | Editorial                                                               |
| Kolek 1994       | Paper not available for review                                          |
| Kolek 1995       | Paper not available for review                                          |
| Kolilekas 2010   | Not a randomised controlled trial or cohort study                       |
| Kolilekas 2013   | Follow-up duration variable                                             |
| Kolilekas 2016   | Editorial                                                               |
| Kondoh 2013      | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Kondoh 2015a     | No relevant outcomes                                                   |
| Kondoh 2016      | Participants overlap with other included study (Natsuiizaka 2014)        |
| Konishi 2012     | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Konishi 2015     | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Kono 2013        | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Kono 2014        | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Kono 2016        | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Koo 2017         | Follow-up duration unclear; author can’t be contacted                   |
| Reference                  | Description                                                                 |
|----------------------------|-----------------------------------------------------------------------------|
| Kopinski 2011              | Not a randomised controlled trial or cohort study                           |
| Kopinski 2011a             | Not a randomised controlled trial or cohort study                           |
| Korthagen 2014             | Follow-up duration variable                                                 |
| Krakówka 1968              | Included participants < 18 years old                                        |
| Kreuter 2014               | Participants overlap with other included study (Kreuter 2016)               |
| Kreuter 2015               | Not a randomised controlled trial or cohort study                           |
| Kreuter 2015a              | Not a randomised controlled trial or cohort study                           |
| Kreuter 2016a              | Not a randomised controlled trial or cohort study                           |
| Kreuter 2016b              | Not a randomised controlled trial or cohort study                           |
| Kreuter 2016c              | Not a randomised controlled trial or cohort study                           |
| Kreuter 2016d              | Not a randomised controlled trial or cohort study                           |
| Kreuter 2017               | Not a randomised controlled trial or cohort study                           |
| Kreuter 2017a              | Not a randomised controlled trial or cohort study                           |
| Krowka 2007                | Follow-up duration < 12 months                                              |
| Kubo 2005                  | Follow-up duration variable; author can’t be contacted                      |
| Kulkarni 2015              | Follow-up duration unclear; author can’t be contacted                       |
| Kulkarni 2016              | Follow-up duration unclear; author can’t be contacted                       |
| Kundu 2014                 | Follow-up duration unclear; author can’t be contacted                       |
| Kunstling 1976             | Review article                                                             |
| Kuse 2016                  | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Kyung 2005                 | Foreign language (translation not available)                                |
| Kärkkäinen 2015            | Follow-up duration unclear; author can’t be contacted                       |
| Kärkkäinen 2016            | Follow-up duration unclear; author can’t be contacted                       |
| Lai 2012a                  | Follow-up duration unclear; author can’t be contacted                       |
| Lama 2003                  | Follow-up duration variable; author can’t be contacted                      |
| Lamas 2011                 | Follow-up duration variable; author contacted                               |
| Lamas 2011a                | Follow-up duration variable; author contacted                               |
| Lancaster 2005             | Follow-up duration variable; author can’t be contacted                      |
| Lancaster 2009             | Not a randomised controlled trial or cohort study                           |
| Lancaster 2015             | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Lancaster 2016             | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Langacker 1981             | Follow-up duration unclear; author can’t be contacted                       |
| Lanser 1984                | Review article                                                             |
| Lavender 2011              | Comment                                                                     |
| Layton 2017                | Mixed disease group                                                         |
| Leceuvre 2016              | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Lederer 2006a              | Follow-up duration < 12 months                                              |
| Lederer 2006b              | Follow-up duration variable; author contacted                               |
| Lee 2005                   | Follow-up duration unclear; author can’t be contacted                       |
| Lee 2009                   | Follow-up duration variable                                                 |
| Lee 2011a                  | Participants overlap with other included study (Ryerson 2014)               |
| Lee 2011b                  | Follow-up duration unclear                                                  |
| Lee 2012a                  | Participants overlap with other included study (Ryerson 2014)               |
| Lee 2012b                  | Follow-up duration unclear                                                  |
| Lee 2012c                  | Follow-up duration variable                                                 |
| Lee 2012d                  | No relevant outcomes                                                       |
| Lee 2013                   | Follow-up duration unclear; author can’t be contacted                       |
| Lee 2013a                  | Participants overlap with other included study (Ryerson 2014)               |
| Lee 2014                   | Follow-up duration variable                                                 |
| Lee 2015a                  | Follow-up duration variable; author can’t be contacted                      |
| Lee 2015b                  | Follow-up duration unclear; author can’t be contacted                       |
| Lee 2015c                  | Follow-up duration variable                                                 |
| Reference        | Note                                                                 |
|------------------|----------------------------------------------------------------------|
| Lee 2016         | Follow-up duration unclear; author can’t be contacted                |
| Lee 2016a        | Follow-up duration unclear; author can’t be contacted                |
| Lee 2016b        | Follow-up duration variable                                           |
| Lee 2017         | Follow-up duration unclear; author can’t be contacted                |
| Lei 1983         | Included participants < 18 years old                                 |
| Lettieri 2006    | Participants overlap with other included study (Fisher 2017)         |
| Lettieri 2006a   | Participants overlap with other included study (Fisher 2017)         |
| Leuchte 2015     | Follow-up duration unclear; author can’t be contacted                |
| Leung 2008       | Mixed disease group                                                  |
| Ley 2011         | Participants overlap with other included studies (Ryerson 2014 and Moua 2016) |
| Ley 2012         | Participants overlap with other included studies (Ryerson 2014 and Moua 2016) |
| Ley 2012a        | Participants overlap with other included studies (Ryerson 2014 and Moua 2016) |
| Ley 2013         | Review article                                                       |
| Ley 2014         | Participants overlap with other included studies (INSPIRE and CAPACITY trials) |
| Ley 2014a        | Participants overlap with other included studies (Ryerson 2014 and Moua 2016) |
| Ley 2015         | Participants overlap with other included studies (INSPIRE and CAPACITY trials) |
| Ley 2015a        | Editorial                                                            |
| Ley 2016         | Not a randomised controlled trial or cohort study                    |
| Li 2010a         | Follow-up duration unclear; author can’t be contacted                |
| Li 2015a         | Follow-up duration variable; author can’t be contacted                |
| Liang 2014       | Not a randomised controlled trial or cohort study                    |
| Lichert 2013     | Paper not available for review                                       |
| Lindell 2007     | Follow-up duration < 12 months                                       |
| Lindell 2011     | Follow-up duration unclear; author can’t be contacted                |
| Lindell 2014     | Follow-up duration unclear; author can’t be contacted                |
| Linden 2006      | Follow-up duration unclear; author can’t be contacted                |
| Loeh 2014        | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Loeh 2015        | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Lok 1999         | Follow-up duration unclear; author can’t be contacted                |
| Louw 1984        | Follow-up duration variable; author can’t be contacted                |
| Lynch 2004       | Follow-up duration unclear                                           |
| Lynch 2004a      | Follow-up duration unclear                                           |
| Lynch 2005       | Follow-up duration unclear                                           |
| Ma 2012          | Follow-up duration unclear                                           |
| Ma 2013          | Follow-up duration unclear; author can’t be contacted                |
| Mackay 2010      | Comment                                                              |
| Makela 2016      | Not a randomised controlled trial or cohort study                    |
| Makiguchi 2016   | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Malagari 2003    | No placebo group                                                     |
| Maldonado 2014   | Follow-up duration unclear                                           |
| Manali 2008      | Follow-up duration variable                                           |
| Mandi 1974       | Not idiopathic pulmonary fibrosis                                     |
| Mannes 1994      | Follow-up duration < 12 months                                       |
| Margaritopoulos 2016 | Comment                                                                |
| Margaritopoulos 2016 | Not a randomised controlled trial or cohort study                    |
| Year  | Study Information                                                                 |
|-------|-----------------------------------------------------------------------------------|
| 2016a | Mart 2012 Follow-up duration unclear; author can’t be contacted                    |
|       | Martinez 2004 Follow-up duration variable; author can’t be contacted               |
|       | Martinez 2005 Follow-up duration variable; author can’t be contacted               |
|       | Martinez 2013 Follow-up duration < 12 months                                       |
|       | Martinez-Moreno 2015 Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
|       | Martinez-Moreno 2016 Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
|       | Marty 1973 Not a randomised controlled trial or cohort study                       |
|       | Mascitelli 2010 Letter                                                             |
|       | Masjedi 2010 Follow-up duration unclear; author can’t be contacted                 |
|       | Mathal 2007 Not a randomised controlled trial or cohort study                       |
|       | Matson 2016 Follow-up duration unclear                                              |
|       | Matsubara 2009 Follow-up duration variable                                          |
|       | Matusiewicz 1993 Follow-up duration variable                                        |
|       | McAllister 2016 Follow-up duration unclear; author can’t be contacted               |
|       | McBurney 2012 Data unclear; author can’t be contacted                                |
|       | McCormack 1991 Participants overlap with other included study (Strand 2014)        |
|       | McCormack 1995 Participants overlap with other included study (Strand 2014)        |
|       | McCormack 1995a Participants overlap with other included study (Strand 2014)       |
|       | Meier-Sydow 1978 Paper not available for review                                     |
|       | Meier-Sydow 1979a Follow-up duration variable                                       |
|       | Meier-Sydow 1986 No relevant outcomes                                              |
|       | Meier-Sydow 1986a No relevant outcomes                                              |
|       | Mejia 2009 Participants overlap with other included study (Richards 2012)          |
|       | Meliconi 1990 Follow-up duration unclear; author can’t be contacted                 |
|       | Meliconi 1990a Follow-up duration unclear; author can’t be contacted                |
|       | Meltzer 2011 Not a randomised controlled trial or cohort study                      |
|       | Meltzer 2011a Not a randomised controlled trial or cohort study                     |
|       | Meyer 2017 Comment                                                                 |
|       | Meyers 2000 Follow-up duration unclear                                              |
|       | Micco 2012 Follow-up duration unclear; author can’t be contacted                    |
|       | Miki 2003 Follow-up duration unclear; author can’t be contacted                     |
|       | Mills 2014 Follow-up duration unclear; author can’t be contacted                    |
|       | Milne 2016 Not a randomised controlled trial or cohort study                        |
|       | Minai 2008 Participants overlap with other included study (Mason 2007)             |
|       | Minai 2012 Not a randomised controlled trial or cohort study                        |
|       | Minegishi 2011 Mixed disease group                                                  |
|       | Minegishi 2011a Mixed disease group                                                 |
|       | Minegishi 2011b Mixed disease group                                                 |
|       | Minegishi 2014 Not idiopathic pulmonary fibrosis                                     |
|       | Minnis 2015 No relevant outcomes                                                   |
|       | Mino 1995 Follow-up duration variable; author can’t be contacted                    |
|       | Mitchell 2011 Not a randomised controlled trial or cohort study                     |
|       | Mitchell 2013 Not a randomised controlled trial or cohort study                     |
|       | Miyake 2006 Not a randomised controlled trial or cohort study                       |
|       | Miyamoto 1992 Foreign language (translation not available)                          |
|       | Mizushima 2010 Follow-up duration unclear; author can’t be contacted                |
|       | Modrykamien 2009 Not a randomised controlled trial or cohort study                  |
|       | Modrykamien 2010 Not a randomised controlled trial or cohort study                  |
|       | Mogulkoc 2001a Follow-up duration unclear; author can’t be contacted                 |
| Study                          | Description                                                                 |
|-------------------------------|-----------------------------------------------------------------------------|
| Mogulkoc 2015                 | Follow-up duration unclear; author can’t be contacted                        |
| Mogulkoc 2016                 | Follow-up duration unclear; author can’t be contacted                        |
| Mohabir 2011                  | Participants overlap with other included study (Su 2011)                    |
| Mohanasundaram 2015           | No relevant outcomes                                                        |
| Molina-Molina 2003            | Mixed disease group                                                         |
| Molina-Molina 2008            | Follow-up duration variable; author can’t be contacted                       |
| Molyneaux 2015                | Participants overlap with other included study (Russell 2016)               |
| Monaghan 2004                 | Participants overlap with other included study (Jacob 2016)                 |
| Montani 2007                  | Comment                                                                     |
| Montesi 2016                  | Follow-up duration unclear; author can’t be contacted                        |
| Moolman 1991                  | Not a randomised controlled trial or cohort study                            |
| Moon 2016                     | No relevant outcomes                                                        |
| Mooney 2012                   | Participants overlap with other included study (Ryerson 2014)               |
| Mooney 2013                   | Participants overlap with other included study (Ryerson 2014)               |
| Moore 2009                    | Editorial                                                                   |
| Moore 2015                    | No relevant outcomes                                                        |
| Mori 2016                     | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Motomura 2012                 | Follow-up duration unclear; author can’t be contacted                        |
| Moua 2011                     | Follow-up duration unclear                                                  |
| Moua 2012                     | Follow-up duration unclear                                                  |
| Moua 2014                     | Participants overlap with other included study (Moua 2016)                  |
| Moua 2014a                    | Follow-up duration variable; author can’t be contacted                       |
| Mudambi 2015                  | Follow-up duration unclear; author can’t be contacted                        |
| Mujakperuo 2013               | Editorial                                                                   |
| Mukae 2002                    | Not a randomised controlled trial or cohort study                            |
| Munteanu 2011                 | Follow-up duration unclear; author can’t be contacted                        |
| Mura 2004                     | Follow-up duration unclear; author can’t be contacted                        |
| Mura 2005                     | Follow-up duration variable; author can’t be contacted                       |
| Mura 2006                     | Follow-up duration variable; author can’t be contacted                       |
| Mura 2006a                    | Not a randomised controlled trial or cohort study                            |
| Nadrous 2005                  | Participants overlap with other included study (Nadrous 2004)                |
| Nadrous 2005a                 | Participants overlap with other included study (Nadrous 2004)                |
| Nadrous 2005b                 | Participants overlap with other included study (Nadrous 2004)                |
| Nagai 1998a                   | Paper not available for review                                              |
| Nagai 1999                    | Follow-up duration unclear; author can’t be contacted                        |
| Nagao 2002                    | Participants overlap with other included study (Hamada 2007)                |
| Nagata 2011                   | Participants overlap with other included study (Akagi 2009)                 |
| Najafizadeh 2011              | Mixed disease group                                                         |
| Nakagawa 2016                 | Follow-up duration unclear; author can’t be contacted                        |
| Nakaya 2011                   | Follow-up duration unclear; author can’t be contacted                        |
| Nakaya 2014                   | Follow-up duration unclear; author can’t be contacted                        |
| Nakayama 2003                 | Not a randomised controlled trial or cohort study                            |
| Nakayama 2011                 | Follow-up duration unclear; author can’t be contacted                        |
| Nakayama 2013                 | Not idiopathic pulmonary fibrosis                                            |
| Nannini 2015                  | Follow-up duration unclear; author can’t be contacted                        |
| Natarajan 2015                | Follow-up duration unclear; author can’t be contacted                        |
| Nathan 2004                   | Follow-up duration variable                                                 |
| Nathan 2007                   | Not a randomised controlled trial or cohort study                            |
| Nathan 2008                   | Follow-up duration variable                                                 |
| Nathan 2008a                  | Not a randomised controlled trial or cohort study                            |
| Reference   | Note                                                                 |
|-------------|----------------------------------------------------------------------|
| Nathan 2010 | Follow-up duration unclear                                           |
| Nathan 2011a| Participants overlap with other included study (Fisher 2017a)        |
| Nathan 2012 | Follow-up duration unclear                                           |
| Nathan 2012a| Participants overlap with other included study (Fisher 2017a)        |
| Nathan 2013 | Follow-up duration unclear                                           |
| Nathan 2015 | Not a randomised controlled trial or cohort study                    |
| Nathan 2015a| Not a randomised controlled trial or cohort study                    |
| Nathan 2015b| Not a randomised controlled trial or cohort study                    |
| Nathan 2015c| Not a randomised controlled trial or cohort study                    |
| Nathan 2015d| Follow-up duration unclear                                           |
| Nathan 2015e| Follow-up duration variable                                          |
| Nathan 2016 | Not a randomised controlled trial or cohort study                    |
| Nathan 2016a| Not a randomised controlled trial or cohort study                    |
| Nathan 2016b| Not a randomised controlled trial or cohort study                    |
| Nathan 2017a| Not a randomised controlled trial or cohort study                    |
| Natsuiaka 2012| Mixed disease group                                                |
| Navaratnam 2010| Follow-up duration variable; author can’t be contacted             |
| Navaratnam 2011| Follow-up duration variable; author can’t be contacted             |
| Navaratnam 2012| Follow-up duration unclear; author can’t be contacted             |
| Navaratnam 2013| Follow-up duration unclear; author can’t be contacted             |
| Navaratnam 2014| Follow-up duration unclear; author can’t be contacted             |
| Navaratnam 2016| Follow-up duration unclear; author can’t be contacted             |
| Navas 2006  | Mixed disease group                                                  |
| Neurohr 2010 | Follow-up duration variable                                          |
| Ng 2016     | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Nicholson 2002| Participants overlap with other included study (Jacob 2016)        |
| Nicol 2015a| Participants overlap with other included study (Nicol 2015)         |
| Nicol 2016 | Follow-up duration unclear; author can’t be contacted             |
| Nicol 2016a| Follow-up duration unclear; author can’t be contacted             |
| Nikaido 2013| Mixed disease group                                                  |
| Nishiyama 2001| Not a randomised controlled trial or cohort study                    |
| Nishiyama 2004| Follow-up duration < 12 months                                       |
| Nishiyama 2007| Not a randomised controlled trial or cohort study                    |
| Nishiyama 2010| Participants overlap with other included study (Kondoh 2010)        |
| Nishiyama 2010a| Participants overlap with other included study (Kondoh 2010)       |
| Nishiyama 2012| Follow-up duration unclear; author can’t be contacted             |
| Nishiyama 2012a| Participants overlap with other included study (Kondoh 2010)        |
| Nishiyama 2016b| Mixed disease group                                                |
| Noble 2010 | Not a randomised controlled trial or cohort study                    |
| Noble 2014 | Not a randomised controlled trial or cohort study                    |
| Noble 2014a| Not a randomised controlled trial or cohort study                    |
| Noble 2015 | Not a randomised controlled trial or cohort study                    |
| Noble 2015a| Not a randomised controlled trial or cohort study                    |
| Noble 2016 | Not a randomised controlled trial or cohort study                    |
| Noble 2016a| Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Noble 2016b| Not a randomised controlled trial or cohort study                    |
| Noble 2016c| Not a randomised controlled trial or cohort study                    |
| Noble 2016d| Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Noth 2013   | Participants overlap with other included study (Oldham 2015)        |
| Noth 2013a | Participants overlap with other included study (Oldham 2015)        |
| Novela 2009 | Follow-up duration < 12 months                                       |
| Nozu 2009   | Mixed disease group                                                  |
| Reference          | Description                                                                 |
|--------------------|-----------------------------------------------------------------------------|
| Nunes 2011         | Follow-up duration variable; author can’t be contacted                       |
| Nunomura 2016      | Follow-up duration variable                                                  |
| O’Dwyer 2013       | Participants overlap with other included study (INSPIRE trial)               |
| Obi-Tabot 2012     | No relevant outcomes                                                         |
| Oda 2014a          | Follow-up duration unclear; author can’t be contacted                        |
| Oda 2016           | Follow-up duration unclear; author can’t be contacted                        |
| Ogawa 2012         | Follow-up duration unclear; author can’t be contacted                        |
| Oh 2012            | Follow-up duration unclear; author can’t be contacted                        |
| Ohkubo 2016        | Follow-up duration unclear; author can’t be contacted                        |
| Ohshimo 2012       | No relevant outcomes                                                         |
| Ohshimo 2013       | Follow-up duration unclear; author can’t be contacted                        |
| Ohshimo 2013a      | Follow-up duration unclear; author can’t be contacted                        |
| Ohshimo 2014       | Follow-up duration unclear; author can’t be contacted                        |
| Ohshimo 2014a      | Follow-up duration unclear; author can’t be contacted                        |
| Ohshimo 2014b      | No relevant outcomes                                                         |
| Ohshimo 2015       | Follow-up duration unclear; author can’t be contacted                        |
| Ohshimo 2015a      | Follow-up duration unclear; author can’t be contacted                        |
| Ohta 2017          | Follow-up duration < 12 months                                               |
| Oishi 2013         | Follow-up duration unclear; author can’t be contacted                        |
| Oishi 2016         | Possible use of antifibrotic agents (nintedanib or pirfenidone); author can’t be contacted |
| Oishi 2016a        | Possible use of antifibrotic agents (nintedanib or pirfenidone); author can’t be contacted |
| Okamoto 2006       | Foreign language (translation not available)                                 |
| Okutan 2012        | Follow-up duration unclear; author can’t be contacted                        |
| Oldham 2015a       | Not a randomised controlled trial or cohort study                            |
| Olson 2007         | Mixed disease group                                                          |
| Olson 2009         | Mixed disease group                                                          |
| Oltmanns 2013      | Included patients receiving antifibrotic agents (nintedanib or pirfenidone)  |
| Oltmanns 2014      | Included patients receiving antifibrotic agents (nintedanib or pirfenidone)  |
| Oltmanns 2014a     | Included patients receiving antifibrotic agents (nintedanib or pirfenidone)  |
| Omori 2015         | Follow-up duration unclear; author can’t be contacted                        |
| Otaola 2011        | Follow-up duration unclear; author can’t be contacted                        |
| Otsuka 2013        | Follow-up duration unclear; author can’t be contacted                        |
| Otsuka 2016        | Follow-up duration unclear; author can’t be contacted                        |
| Ozawa 2009         | Included patients receiving antifibrotic agents (nintedanib or pirfenidone)  |
| Paik 2012          | Follow-up duration unclear; author can’t be contacted                        |
| Palwatwichai 2000  | Follow-up duration unclear; author can’t be contacted                        |
| Pannu 2015         | Follow-up duration unclear; author can’t be contacted                        |
| Papali 2010        | Follow-up duration unclear                                                  |
| Papiris 1997       | Follow-up duration unclear; author can’t be contacted                        |
| Papiris 2015       | Follow-up duration variable                                                  |
| Parambil 2005      | Not a randomised controlled trial or cohort study                            |
| Parfrey 2012       | Included patients receiving antifibrotic agents (nintedanib or pirfenidone)  |
| Parfrey 2013       | Included patients receiving antifibrotic agents (nintedanib or pirfenidone)  |
| Parfrey 2014       | Included patients receiving antifibrotic agents (nintedanib or pirfenidone)  |
| Park 1999          | Not a randomised controlled trial or cohort study                            |
| Park 2004          | Foreign language (translation not available)                                 |
| Park 2007          | Participants overlap with other included study (Song 2011)                   |
| Park 2012          | Follow-up duration unclear                                                  |
| Park 2012a         | Follow-up duration variable; author can’t be contacted                       |
| Park 2012b         | Follow-up duration variable; author can’t be contacted                       |
| Author(s)       | Notes                                                                                             |
|----------------|--------------------------------------------------------------------------------------------------|
| Park 2016      | Participants overlap with other included study (Song 2011)                                        |
| Park 2016a     | Follow-up duration unclear; author can't be contacted                                             |
| Parra 2006     | Not a randomised controlled trial or cohort study                                                 |
| Parra 2007     | Follow-up duration unclear; author can't be contacted                                             |
| Parra 2007a    | Follow-up duration unclear; author can't be contacted                                             |
| Parra 2008     | Follow-up duration unclear; author can’t be contacted                                             |
| Parra 2010     | Follow-up duration variable; author can’t be contacted                                             |
| Parra 2012     | Participants overlap with other included study (Soares 2015)                                      |
| Parra 2012a    | Participants overlap with other included study (Soares 2015)                                      |
| Parra 2012b    | Participants overlap with other included study (Soares 2015)                                      |
| Parra 2012c    | Follow-up duration variable; author can’t be contacted                                             |
| Parra 2013     | Participants overlap with other included study (Soares 2015)                                      |
| Parra 2013a    | Participants overlap with other included study (Soares 2015)                                      |
| Parra 2014     | Participants overlap with other included study (Soares 2015)                                      |
| Paterniti 2017 | Not a randomised controlled trial or cohort study                                                 |
| Patterson 2017 | Data unclear; author cant be contacted                                                             |
| Peelen 2010    | Follow-up duration unclear; author can’t be contacted                                              |
| Peljto 2013    | Participants overlap with other included study (Oldham 2015)                                       |
| Peng 2008      | Follow-up duration unclear                                                                       |
| Pereira 2006   | Participants overlap with other included study (Soares 2015)                                      |
| Perez-Padilla 1993 | Participants overlap with other included study (Selman 1998)                                    |
| Peris 2011     | Follow-up duration unclear; author can’t be contacted                                              |
| Peters 1993    | Follow-up duration variable                                                                       |
| Pires 2011     | Follow-up duration unclear; author can’t be contacted                                              |
| Pitsiou 2007   | Follow-up duration unclear; author can’t be contacted                                              |
| Pittrow 2014   | Not a randomised controlled trial or cohort study                                                 |
| Pohl 1993      | Diagnosis unclear; author can’t be contacted                                                       |
| Polonski 1994  | Paper not available for review                                                                   |
| Polonski 1995  | Foreign language (translation not available)                                                       |
| Polonski 1995a | Paper not available for review                                                                   |
| Poor 2010      | Follow-up duration unclear; author can’t be contacted                                              |
| Portillo 2016  | Mixed disease group                                                                               |
| Prasse 2003    | Not a randomised controlled trial or cohort study                                                 |
| Prasse 2008    | Paper not available for review                                                                   |
| Prasse 2012    | Follow-up duration unclear                                                                       |
| Prasse 2015    | Follow-up duration unclear; author can’t be contacted                                              |
| Pritchett 2012 | Not a randomised controlled trial or cohort study                                                 |
| Probst 2011    | Participants overlap with other included study (Prasse 2009)                                      |
| Probst 2012    | Participants overlap with other included study (Prasse 2009)                                      |
| Pujols 2004    | Follow-up duration variable; author can’t be contacted                                              |
| Puthiyaveettil 2014 | Included patients receiving antifibrotic agents (nintedanib or pirfenidone)                   |
| Putman 2016    | Not idiopathic pulmonary fibrosis                                                                |
| Quadrelli 2010 | Follow-up duration unclear; author can’t be contacted                                              |
| Quinn 2002     | Not a randomised controlled trial or cohort study                                                 |
| Rad 2015       | Mixed disease group                                                                               |
| Raghu 1991     | No placebo group                                                                                  |
| Raghu 1999     | Included patients receiving antifibrotic agents (nintedanib or pirfenidone)                       |
| Raghu 2000     | Follow-up duration < 12 months                                                                    |
| Raghu 2001     | Follow-up duration < 12 months                                                                    |
| Raghu 2003     | Follow-up duration variable; author can’t be contacted                                              |
| Raghu 2004     | Follow-up duration variable; author can’t be contacted                                              |
| Raghu 2004a    | Follow-up duration unclear; author can’t be contacted                                              |
| Reference                  | Description                                                                 |
|---------------------------|-----------------------------------------------------------------------------|
| Raghu 2006a               | Not a randomised controlled trial or cohort study                            |
| Raghu 2007                | Follow-up duration < 12 months                                               |
| Raghu 2008                | Follow-up duration < 12 months                                               |
| Raghu 2012a               | Follow-up duration < 12 months                                               |
| Raghu 2012b               | Follow-up duration < 12 months                                               |
| Raghu 2012c               | Follow-up duration < 12 months                                               |
| Raghu 2013b               | Participants overlap with other included study (Collins 2015)               |
| Raghu 2014b               | Follow-up duration variable; author can’t be contacted                       |
| Raghu 2016                | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Raghu 2016a               | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Raghu 2016b               | Review article                                                              |
| Raghu 2016c               | Participants overlap with other included study (Collins 2015)               |
| Raghu 2016d               | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Rajasekaran 2001          | Not a randomised controlled trial or cohort study                            |
| Rathnapala 2016           | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Rathnapala 2016a          | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Rathnapala 2016b          | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Ravaglia 2013             | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Rebeck 1982               | Mixed disease group                                                          |
| Redfern 2016              | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Redondo 2014              | Not a randomised controlled trial or cohort study                            |
| Redondo 2014a             | Participants overlap with other included study (Soares Pires 2013)           |
| Reed 2006                 | Mixed disease group                                                          |
| Reichner 2004             | Paper not available for review                                               |
| Relf 2015                 | Follow-up duration unclear; author can’t be contacted                        |
| Ren 2014                  | Follow-up duration unclear; author can’t be contacted                        |
| Renzoni 1997              | Follow-up duration unclear; author can’t be contacted                        |
| Riario Sforza 2008        | Editorial                                                                    |
| Ribeiro Neto 2013         | Letter                                                                       |
| Richards 2011             | Follow-up duration unclear; author can’t be contacted                        |
| Richards 2012a            | Participants overlap with other included study (Richards 2012)               |
| Richeldi 2004             | Letter                                                                       |
| Richeldi 2012             | Participants overlap with other included studies (Ryerson 2014 and Moua 2016) |
| Richeldi 2013             | Not a randomised controlled trial or cohort study                            |
| Richeldi 2014b            | Study protocol                                                               |
| Richeldi 2015             | Not a randomised controlled trial or cohort study                            |
| Richeldi 2015a            | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Richeldi 2015b            | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Richeldi 2016             | Not a randomised controlled trial or cohort study                            |
| Richeldi 2016a            | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Riddell 2013              | Follow-up duration unclear; author can’t be contacted                        |
| Riddell 2013a             | Follow-up duration unclear; author can’t be contacted                        |
| Riddell 2014              | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Riha 2002                 | Follow-up duration variable; author can’t be contacted                       |
| Rivera-Lebron 2012        | Follow-up duration unclear; author can’t be contacted                        |
| Rivera-Lebron 2013        | Follow-up duration unclear; author can’t be contacted                        |
| Roldán 2016               | Not a randomised controlled trial or cohort study                            |
| Romagnoli 2012            | Follow-up duration unclear; author can’t be contacted                        |
| Romei 2012                | Follow-up duration unclear                                                  |
| Romei 2015                | Mixed disease group                                                          |
| Rooney 2016               | Follow-up duration < 12 months                                               |
| Reference          | Status                                                                 |
|--------------------|-------------------------------------------------------------------------|
| Rudd 1981          | Mixed disease group                                                     |
| Rufino 2011        | Follow-up duration unclear; author can’t be contacted                   |
| Rusanov 2012       | No relevant outcomes                                                   |
| Rush 2016          | Follow-up duration unclear; author can’t be contacted                   |
| Russell 2013       | Not a randomised controlled trial or cohort study                       |
| Ryerson 2010       | Not a randomised controlled trial or cohort study                       |
| Ryerson 2011       | Follow-up duration unclear; participants overlap with other included study (Ryerson 2014) |
| Ryerson 2011a      | Follow-up duration unclear; participants overlap with other included study (Ryerson 2014) |
| Ryerson 2012       | Participants overlap with other included studies (Ryerson 2014 and Moua 2016) |
| Ryerson 2013       | Participants overlap with other included studies (Ryerson 2014 and Moua 2016) |
| Ryerson 2013a      | Participants overlap with other included study (Ryerson 2014)           |
| Ryerson 2015       | Not idiopathic pulmonary fibrosis                                       |
| Saini 2013         | Not a randomised controlled trial or cohort study                       |
| Saini 2015         | Diagnosis unclear; author can’t be contacted                            |
| Saito 2011         | Participants overlap with other included study (Kurashima 2010)         |
| Sakamoto 2011      | Follow-up duration unclear                                              |
| Sakamoto 2012      | Follow-up duration unclear                                              |
| Sakamoto 2015      | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Salinas 2012       | Foreign language (translation not available)                            |
| Salinas 2014       | Foreign language (translation not available)                            |
| Salisbury 2015     | Participants overlap with other included studies (Jacob 2016 and Gay 1998) |
| Salisbury 2016     | Participants overlap with other included study (PANTHER-IPF)            |
| Salisbury 2016a    | Participants overlap with other included studies (Jacob 2016 and Gay 1998) |
| Santana 2008       | Letter                                                                  |
| Santos 2016        | Mixed disease group                                                     |
| Saravanan 2003     | Letter                                                                  |
| Sarwar 2016        | Not a randomised controlled trial or cohort study                       |
| Sato 2013          | Follow-up duration unclear; author can’t be contacted                   |
| Sato 2013a         | Follow-up duration unclear; author can’t be contacted                   |
| Sato 2016a         | Follow-up duration variable; author can’t be contacted                  |
| Sato 2006          | Mixed disease group                                                     |
| Scadding 1967      | Not idiopathic pulmonary fibrosis                                       |
| Scalori 2014       | Study protocol                                                          |
| Schachna 2006      | Follow-up duration variable                                              |
| Schafer 2013       | No relevant outcomes                                                   |
| Schafer 2012       | Follow-up duration unclear                                               |
| Schilde 2011       | Follow-up duration unclear; author can’t be contacted                   |
| Schmidt 1987       | No relevant outcomes                                                   |
| Schmidt 2011a      | Participants overlap with other included studies (Jacob 2016 and Gay 1998) |
| Schmidt 2011b      | Not a randomised controlled trial or cohort study                       |
| Schmidt 2014       | Participants overlap with other included studies (Jacob 2016 and Gay 1998) |
| Schupp 2016        | Participants overlap with other included study (Schupp 2015)            |
| Schwartz 1991      | Not a randomised controlled trial or cohort study                       |
| Schwartz 1994      | Follow-up duration variable; author can’t be contacted                  |
| Study                                      | Comments                                                                 |
|--------------------------------------------|--------------------------------------------------------------------------|
| Schwartz 1994a                            | Follow-up duration unclear; author can’t be contacted                    |
| Scientific Committee 2009                  | Foreign language - translation not available                             |
| Scott 2013                                 | Follow-up duration < 12 months                                           |
| Sengul 2009                                | Foreign language (translation not available)                             |
| Serban 2013                                | Not idiopathic pulmonary fibrosis                                        |
| Sestini 2006                               | Mixed disease group                                                     |
| Setoguchi 2009                             | Paper not available for review                                           |
| Shah 1972                                  | Not idiopathic pulmonary fibrosis                                        |
| Shah 2014                                  | Follow-up duration variable                                              |
| Shaker 2013                                | Follow-up duration unclear; author can’t be contacted                    |
| Sharif 2016                                | Not a randomised controlled trial or cohort study                        |
| Sharif 2016a                               | Follow-up duration unclear; author can’t be contacted                    |
| Sharp 2016                                 | Not a randomised controlled trial or cohort study                        |
| Sharp 2016a                                | Follow-up duration unclear                                              |
| Sharp 2017                                 | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Sheikh 2017                               | Follow-up duration unclear                                              |
| Shin 2015                                  | Follow-up duration unclear                                              |
| Shin 2016                                  | Follow-up duration unclear                                              |
| Shino 2012                                 | Follow-up duration unclear; author can’t be contacted                    |
| Shioya 2013                                | Follow-up duration unclear; author can’t be contacted                    |
| Shiraki 2015                               | Follow-up duration unclear; author can’t be contacted                    |
| Shiratori 2017                             | Follow-up duration unclear; author can’t be contacted                    |
| Sholl 2010                                 | Mixed disease group                                                     |
| Shorr 2002                                 | Follow-up duration unclear                                              |
| Shulgina 2011                              | Mixed disease group                                                     |
| Shulgina 2013                              | Mixed disease group                                                     |
| Silva 2007                                 | Mixed disease group                                                     |
| Silva 2008                                 | Participants overlap with other included study (Jacob 2016)              |
| Simon-Blancal 2012                         | Follow-up duration unclear; author can’t be contacted                    |
| Sköld 2016                                 | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Smadja 2013                                | Follow-up duration variable; author can’t be contacted                   |
| Smadja 2014                                | Not a randomised controlled trial or cohort study                        |
| Smith 1990                                 | Follow-up duration variable; author can’t be contacted                   |
| Snell 2016                                 | Not a randomised controlled trial or cohort study                        |
| Soares 2013a                               | Follow-up duration unclear; author can’t be contacted                    |
| Sokai 2014                                 | Follow-up duration < 12 months                                           |
| Sokai 2015                                 | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Sokai 2015a                                | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Sokai 2017                                 | Follow-up duration unclear; author can’t be contacted                    |
| Son 2010                                   | Follow-up duration unclear; author can’t be contacted                    |
| Son 2015                                   | Follow-up duration unclear; author can’t be contacted                    |
| Song 2009                                  | Participants overlap with other included study (Song 2011)               |
| Song 2009a                                 | Follow-up duration variable                                              |
| Song 2010                                  | Participants overlap with other included study (Song 2011)               |
| Song 2011a                                 | Participants overlap with other included study (Song 2011)               |
| Song 2013                                  | Participants overlap with other included study (Song 2011)               |
| Song 2015                                  | No relevant outcomes                                                    |
| Spencer 2011                               | Follow-up duration unclear; author can’t be contacted                    |
| Speranskaya 2015                           | Paper not available for review                                           |
| Stauffer 2016                              | Participants overlap with other included study (Raghu 2014)               |
| Steffensen 1992                            | Not a randomised controlled trial or cohort study                        |
| Author            | Comment                                                                 |
|------------------|--------------------------------------------------------------------------|
| Takenaka 2014    | Participants overlap with other included studies (Oldham 2015 and Ryerson 2014) |
| Sturani 1988     | Paper not available for review                                           |
| Sturani 1990     | Foreign language (translation not available)                              |
| Subhash 2004     | Follow-up duration variable; author can’t be contacted                   |
| Sugino 2010      | Foreign language (translation not available)                              |
| Sugino 2013      | Not idiopathic pulmonary fibrosis                                         |
| Sugino 2014      | Not a randomised controlled trial or cohort study                         |
| Sugino 2014a     | Follow-up duration unclear; author can’t be contacted                     |
| Sugino 2015      | Follow-up duration unclear; author can’t be contacted                     |
| Sugino 2015a     | Follow-up duration unclear; author can’t be contacted                     |
| Suissa 2015      | Comment                                                                   |
| Sumikawa 2008    | Participants overlap with other included studies (Kondoh 2005, Kondoh 2010 and Fujimoto 2012) |
| Sumikawa 2014    | Participants overlap with other included studies (Kondoh 2005, Kondoh 2010 and Fujimoto 2012) |
| Sun 2016         | Paper not available for review                                            |
| Suraj 2016       | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Sverzellati 2011 | Mixed disease group                                                       |
| Sverzellati 2017 | Editorial                                                                  |
| Swift 2013       | Mixed disease group                                                       |
| Swigris 2009     | Follow-up duration variable; author contacted                             |
| Swigris 2011     | Follow-up duration variable; author contacted                             |
| Swigris 2012     | Not a randomised controlled trial or cohort study                         |
| Swigris 2013     | Letter                                                                    |
| Tabuena 2005     | Follow-up duration unclear; author can’t be contacted                     |
| Tachibana 2016   | Follow-up duration unclear; author can’t be contacted                     |
| Tajiri 2003      | Not a randomised controlled trial or cohort study                         |
| Tajiri 2015      | Possible use of antifibrotic agents (nintedanib or pirfenidone); author can’t be contacted |
| Takada 2013      | Mixed disease group                                                       |
| Takada 2014      | Mixed disease group                                                       |
| Takahashi 2006   | Follow-up duration unclear; author can’t be contacted                     |
| Takahashi 2012   | Not a randomised controlled trial or cohort study                         |
| Takaiwa 2014     | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Takei 2016       | Mixed disease group                                                       |
| Takenaka 1999    | Mixed disease group                                                       |
| Takoi 2012       | Mixed disease group                                                       |
| Taniguchi 2011a  | Follow-up duration unclear                                               |
| Taniguchi 2012   | Follow-up duration < 12 months                                            |
| Taniguchi 2014   | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Tanizawa 2015    | Follow-up duration < 12 months                                            |
| Tcherkian 2011   | Follow-up duration variable; author can’t be contacted                    |
| ten Klooster 2011| Follow-up duration unclear; author can’t be contacted                     |
| ten Klooster 2012| Follow-up duration unclear; author can’t be contacted                     |
| ten Klooster 2013| Follow-up duration unclear; author can’t be contacted                     |
| ten Klooster 2015a| Follow-up duration unclear; author can’t be contacted                     |
| Teramachi 2017   | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Terriff 1992     | Follow-up duration variable; author can’t be contacted                    |
| Thabut 2003      | Follow-up duration unclear; author can’t be contacted                     |
| Author Year | Details |
|-------------|---------|
| Thomeer 2004 | Follow-up duration variable; author can’t be contacted |
| Titto 2005 | Follow-up duration unclear; author can’t be contacted |
| Titto 2006 | Follow-up duration unclear; author can’t be contacted |
| Todd 2011 | Mixed disease group |
| Tokura 2009 | Foreign language (translation not available) |
| Tomassetti 2010 | Follow-up duration variable; author can’t be contacted |
| Tomassetti 2012 | Follow-up duration unclear; author can’t be contacted |
| Tomassetti 2013 | Follow-up duration variable; author can’t be contacted |
| Tomassetti 2015 | Follow-up duration variable; author can’t be contacted |
| Tomic 2014 | Follow-up duration unclear; author can’t be contacted |
| Tomic 2015 | Follow-up duration unclear; author can’t be contacted |
| Tomioka 2003 | Participants overlap with other included study (Tomioka 2007) |
| Tomioka 2005 | Participants overlap with other included study (Tomioka 2007) |
| Tomioka 2007a | Follow-up duration variable; author can’t be contacted |
| Tossier 2016 | Follow-up duration unclear; author can’t be contacted |
| Travis 2000 | Follow-up duration variable; author can’t be contacted |
| Triantafillidou 2011 | Follow-up duration variable; author can’t be contacted |
| Triantafillidou 2011a | Follow-up duration unclear; author can’t be contacted |
| Triantafillidou 2013 | Follow-up duration variable; author can’t be contacted |
| Troy 2014 | Not a randomised controlled trial or cohort study |
| Troy 2014a | Not a randomised controlled trial or cohort study |
| Trujillo 2010 | Follow-up duration unclear; author can’t be contacted |
| Tsuboi 2006 | Not a randomised controlled trial or cohort study |
| Tsuchida 2011 | Follow-up duration unclear; author can’t be contacted |
| Tsuchiya 2010 | Foreign language (translation not available) |
| Tsuchiya 2015 | Not a randomised controlled trial or cohort study |
| Tsukamoto 2000 | Follow-up duration unclear; author can’t be contacted |
| Tsushima 2010 | Not idiopathic pulmonary fibrosis |
| Tsutsumi 2015 | Follow-up duration unclear; author can’t be contacted |
| Tukiainen 1979 | Foreign language (translation not available) |
| Tukiainen 1983 | Mixed disease group |
| Turner-Warwick 1980 | Mixed disease group |
| Turner-Warwick 1986 | Review article |
| Turner-Warwick 1987 | Mixed disease group |
| Tzortzaki 2007 | No placebo group |
| Tzouvelekis 2011 | Participants overlap with other included studies (Tzouvelekis 2013) |
| Tzouvelekis 2011a | Participants overlap with other included studies (Tzouvelekis 2013) |
| Tzouvelekis 2011b | Participants overlap with other included studies (Tzouvelekis 2013) |
| Tzouvelekis 2013a | Comment |
| Tzouvelekis 2013b | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Tzouvelekis 2014 | Follow-up duration unclear |
| Uehara 2016 | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Umeda 2009 | Participants overlap with other included study (Umeda 2015) |
| Umeda 2012 | Follow-up duration unclear |
| Umeda 2013 | Follow-up duration unclear; author can’t be contacted |
| Undurraga 1998 | Follow-up duration variable |
| Usui 2011 | Mixed disease group |
| Usui 2013 | Mixed disease group |
| Vainshelboim 2016 | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Vainshelboim 2016a | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Valenzuela 2015 | Follow-up duration unclear; author can’t be contacted |
| Valenzuela 2015 | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Author          | Included patients receiving antifibrotic agents (nintedanib or pirfenidone)                                                                 |
|-----------------|----------------------------------------------------------------------------------------------------------------------------------|
| Valeyre 2014    | Mixed disease group                                                                                                             |
| Van Der Aar 2016| Follow-up duration < 12 months                                                                                                   |
| Van der Plas 2011| Follow-up duration variable                                                                                                      |
| Van der Plas 2014| Follow-up duration variable                                                                                                      |
| Van der Velden 2016| No relevant outcomes                                                                                                            |
| van der Vis 2016| Follow-up duration unclear                                                                                                       |
| van Oortegem 1994| Data unclear; author can't be contacted                                                                                           |
| Varela 2016     | Follow-up duration unclear; author can't be contacted                                                                             |
| Varney 2001     | Follow-up duration < 12 months                                                                                                   |
| Varney 2002     | Follow-up duration < 12 months                                                                                                   |
| Varney 2008     | Mixed disease group                                                                                                              |
| Vasakova 2007   | Follow-up duration unclear                                                                                                       |
| Vasakova 2016   | Participants overlap with other included study (Zurkova 2016)                                                                      |
| Vasakova 2016a  | Participants overlap with other included study (Zurkova 2016)                                                                      |
| Vedel-Krogh 2015| Diagnosis unclear; author can't be contacted                                                                                      |
| Veeraraghavan 2003| Follow-up duration variable                                                                                                      |
| Venuta 1993     | Follow-up duration unclear; author can't be contacted                                                                             |
| Vercauteren 2014| Follow-up duration unclear; author can't be contacted                                                                             |
| Vercauteren 2015| Follow-up duration unclear; author can't be contacted                                                                             |
| Vial Dupuy 2012 | Mixed disease group                                                                                                              |
| Vial-Dupuy 2011 | Mixed disease group                                                                                                              |
| Vial-Dupuy 2011a| Mixed disease group                                                                                                              |
| Vianello 2014   | Follow-up duration unclear; author can’t be contacted                                                                             |
| Vij 2011        | Participants overlap with other included study (Oldham 2015)                                                                      |
| Villanueva Bueno 2016| Included patients receiving antifibrotic agents (nintedanib or pirfenidone)                                                      |
| Vitale 2014     | Patient age unclear; author can’t be contacted                                                                                  |
| Voltolini 2013  | Mixed disease group                                                                                                              |
| Vuga 2014       | Participants overlap with other included study (Richards 2012)                                                                    |
| Wacker 2014     | Not a randomised controlled trial or cohort study                                                                                 |
| Waisberg 2012   | Participants overlap with other included study (Soares 2015)                                                                      |
| Wakamatsu 2016  | Included patients receiving antifibrotic agents (nintedanib or pirfenidone)                                                      |
| Walsh 2012      | Follow-up duration unclear; author can’t be contacted                                                                             |
| Walter 2013     | Follow-up duration unclear; author can’t be contacted                                                                             |
| Wang 2014       | Review article                                                                                                                   |
| Warrington 2010 | Diagnosis and follow-up duration unclear; author can’t be contacted                                                              |
| Watanabe 2008   | Follow-up duration unclear; author can’t be contacted                                                                             |
| Watanabe 2011   | Foreign language (translation not available)                                                                                      |
| Watanabe 2012   | Not idiopathic pulmonary fibrosis                                                                                                |
| Watanabe 2013   | Follow-up duration variable                                                                                                       |
| Watanabe 2014   | Follow-up duration variable                                                                                                       |
| Watters 1987    | Follow-up duration variable; author can’t be contacted                                                                             |
| Wei 2004        | Review article                                                                                                                   |
| Wei 2013        | Review article                                                                                                                   |
| Weinstein 2014  | Included patients receiving antifibrotic agents (nintedanib or pirfenidone)                                                      |
| Weiss 2009      | Mixed disease group                                                                                                              |
| Wells 1993      | Participants overlap with other included study (Jacob 2016)                                                                      |
| Wells 1993a     | Participants overlap with other included study (Jacob 2016)                                                                      |
| Wells 1993b     | Mixed disease group                                                                                                              |
| Wells 1994      | Follow-up duration variable; author can’t be contacted                                                                             |
| Wells 1996      | Mixed disease group                                                                                                              |
| Wells 2003      | Participants overlap with other included study (Jacob 2016)                                                                      |
| Author          | Description                                                                 |
|-----------------|-----------------------------------------------------------------------------|
| Wells 2005      | Follow-up duration variable; author can’t be contacted                       |
| Wells 2016      | Not a randomised controlled trial or cohort study                            |
| Wesolowski 2000 | Follow-up duration uncertain; author can’t be contacted                      |
| Westhoff 2014   | Not idiopathic pulmonary fibrosis                                            |
| White 2012      | Not idiopathic pulmonary fibrosis                                            |
| White 2015      | Not a randomised controlled trial or cohort study                            |
| White 2016      | Follow-up duration < 12 months                                              |
| White 2016a     | Not a randomised controlled trial or cohort study                            |
| Wiertz 2016     | Follow-up duration < 12 months                                              |
| Wijsenbeek 2013 | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Wijsenbeek 2015 | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Wilkie 2013     | Participants overlap with other included study (Wilkie 2012)                 |
| Winget 1997     | Follow-up duration < 12 months                                              |
| Winterbauer 2000| No placebo group                                                            |
| Winterbottom 2014| Follow-up duration uncertain; author can’t be contacted                     |
| Won 2011        | Mixed disease group                                                          |
| Wong 2013       | Not a randomised controlled trial or cohort study                            |
| Woo 2003        | Foreign language (translation not available)                                 |
| Wu 2005         | Follow-up duration < 12 months                                              |
| Wu 2013         | Not a randomised controlled trial or cohort study                            |
| Wu 2015a        | Not a randomised controlled trial or cohort study                            |
| Wu 2016         | Data unclear; author can’t be contacted                                       |
| Wuyts 2016      | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Wyeth-Ayerst 2004 | Follow-up duration < 12 months                     |
| Xaubet 2001     | Follow-up duration uncertain; author can’t be contacted                      |
| Xaubet 2003     | Follow-up duration variable; author can’t be contacted                      |
| Xaubet 2010     | Follow-up duration uncertain; author can’t be contacted                      |
| Xu 2011         | Follow-up duration variable; author can’t be contacted                      |
| Xue 2011        | Follow-up duration uncertain                                                |
| Yagi 2014       | Follow-up duration < 12 months                                              |
| Yagihashi 2015  | Participant overlap with other included studies (IPF Net trials)             |
| Yamada 2003     | Paper not available for review                                              |
| Yamaguchi 1974  | Foreign language (translation not available)                                 |
| Yamaguchi 2017  | Follow-up duration unclear                                                  |
| Yamauchi 2011   | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Yamauchi 2011a  | Follow-up duration < 12 months                                              |
| Yamauchi 2014   | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Yamauchi 2015   | Participants overlap with other included studies (Kondoh 2005, Kondoh 2010 and Fujimoto 2012) |
| Yamauchi 2016   | Participants overlap with other included studies (Kondoh 2005, Kondoh 2010 and Fujimoto 2012) |
| Yamazaki 2016   | Follow-up duration < 12 months                                              |
| Yang 2015       | Follow-up duration uncertain; author can’t be contacted                      |
| Yano 2011       | Mixed disease group                                                          |
| Yasui 2016      | Mixed disease group                                                          |
| Yazaki 2016     | No relevant outcomes                                                        |
| Ye 2014         | Paper not available for review                                              |
| Yokoo 2013      | Follow-up duration uncertain; author can’t be contacted                      |
| Yokoo 2013a     | Follow-up duration uncertain; author can’t be contacted                      |
| Yokoyama 1998   | Follow-up duration uncertain; author can’t be contacted                      |
| Yokoyama 2010   | Follow-up duration < 12 months                                              |
| Yong 2001       | Follow-up duration < 12 months                                              |
| Reference | Description |
|-----------|-------------|
| Yoon 2016 | Included patients receiving antifibrotic agents (nintedanib or pirfenidone) |
| Young 2006 | Follow-up duration variable; author contacted |
| Young 2017 | Mixed disease group |
| Yu 2015 | Follow-up duration variable |
| Yu 2016a | Follow-up duration variable |
| Yukiko 2013 | Mixed disease group |
| Yukiko 2014 | Mixed disease group |
| Zappala 2010 | Participants overlap with other included study (Jacob 2016) |
| Zhang 2010 | Not a randomised controlled trial or cohort study |
| Zhang 2011 | Diagnosis unclear; author can’t be contacted |
| Zhang 2011a | Follow-up duration unclear; author can’t be contacted |
| Zhang 2016a | Not idiopathic pulmonary fibrosis |
| Zhong 2012 | Follow-up duration unclear |
| Ziegenhagen 1998 | Not a randomised controlled trial or cohort study |
| Ziesche 1997 | Patient age unclear; author can’t be contacted |
| Ziesche 1999 | No placebo group |
| Zimmermann 2011 | Mixed disease group |
| Zisman 2000 | Data unclear; author can’t be contacted |
| Zisman 2009 | Follow-up duration variable |
| Zisman 2010 | Follow-up duration < 12 months |
| Zompatori 1996 | Foreign language (translation not available) |
| Zompatori 1997 | Foreign language (translation not available) |
| Zotti 2003 | Follow-up duration < 12 months |

**Part B: References of Excluded Studies**

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### S5. Characteristics of included studies

**Agustí 1993 [22]**

| Study Design | Type of Study: prospective cohort  
Trial Design: single centre  
Country/ies: Spain |
|--------------|----------------------------------------------------------|
| Study Duration | 1 year |
| Participants | Number: 10  
Definition of diagnosis: open lung biopsy in two participants and by the Turner-Warwick criteria (clinical criteria: 1) widespread, persistent bilateral radiographic shadowing; 2) widespread, persisting crackles; and 3) exclusion of those patients in whom an external fibrogenic agent could be implicated, and those with positive plasma precipitins) in the rest  
Age: mean 66 years (standard deviation: 10)  
Gender: 20% male  
Ethnicity: not stated  
Smoking status: not stated  
Use of home oxygen therapy: not stated  
Time since diagnosis: not stated  
Percentage of patients with surgical lung biopsy: 20%  
Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated  
Lung function results (% predicted): mean baseline FVC 47% (standard deviation: 11); mean baseline DLCO 41% (standard deviation: 12)  
Resting oxyhaemoglobin saturation: not stated  
6-minute walk distance: not stated  
Symptom assessment: not stated  
Use of systemic corticosteroid therapy: 90%  
Use of other therapy: aerosolised ribavirin 100% |
| Outcomes | Proportion of mortality |
| Notes | Funding source: Hubber laboratory |

**Agustí 1994 [23]**

| Study Design | Type of Study: prospective cohort  
Trial Design: single centre  
Country/ies: Spain |
|--------------|----------------------------------------------------------|
| Study Duration | 3 years |
| Participants | Number: 27  
Definition of diagnosis: open lung biopsy and clinical criteria of IPF described by Turner-Warwick et al: 1) widespread, persistent bilateral radiographic shadowing; 2) widespread, persisting crackles; and 3) exclusion of those patients in whom an external fibrogenic agent could be implicated, and those with positive plasma precipitins  
Age: mean 55 years (standard deviation: 14)  
Gender: 63% male  
Ethnicity: not stated  
Smoking status: 52% ever smokers  
Use of home oxygen therapy: not stated  
Time since diagnosis: not stated  
Percentage of patients with surgical lung biopsy: 52%  
Percentage of patients with definite usual interstitial pneumonia |
| Notes | Funding source: Hubber laboratory |
patterns on HRCT: not stated

- Lung function results (% predicted): mean FVC 62% (standard deviation: 14); mean DLCO 55% (standard deviation: 14)
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: not assessed
- Use of systemic corticosteroid therapy: not stated
- Use of other therapy: not stated

### Outcomes
- Proportion of mortality
- Change in forced vital capacity

### Notes
- Funding source: not stated

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**Akagi 2009 [24]**

| Study Design | Type of Study: retrospective cohort
|             | Trial Design: single centre
|             | Country/ies: Japan

| Study Duration | 1 year

| Participants | Number: 59 (33 IPF)
|             | Definition of diagnosis: the 2002 American Thoracic Society/European Respiratory Society International Multidisciplinary Consensus Classification of the Idiopathic Interstitial Pneumonias Statement
|             | Age: mean 66.5 years (standard deviation: 9.2)
|             | Gender: 66% male
|             | Ethnicity: not stated
|             | Smoking status: 76% ever smoker
|             | Use of home oxygen therapy: not stated
|             | Time since diagnosis: not stated
|             | Percentage of patients with surgical lung biopsy: 26%
|             | Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
|             | Lung function results (% predicted): mean FVC 72.8% (standard deviation: 19.4); mean DLCO 60.7% (standard deviation: 19.8)
|             | Resting oxyhaemoglobin saturation: Not stated
|             | 6-minute walk distance: not stated
|             | Symptom assessment: not assessed
|             | Use of systemic corticosteroid therapy: 45%
|             | Use of other therapy: not stated

| Outcomes | Proportion of mortality

| Notes | Funding source: not stated

Author provided additional data for clarification of study duration and survival/mortality.

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**Alhamad 2008 [25]**

| Study Design | Type of Study: retrospective cohort
|             | Trial Design: single centre
|             | Country/ies: Saudi Arabia

| Study Duration | At least 1 year, mean 4.2 years

| Participants | Number: 61
|             | Definition of diagnosis: the 2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment International Consensus Statement
|             | Age: mean 54.7 years (standard deviation: 15.2)
- Gender: 49% male
- Ethnicity: not stated
- Smoking status: not stated
- Use of home oxygen therapy: 21%
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: 73%
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results (% predicted): mean FVC 61.5% (standard deviation: 25.2) in surgical biopsy group, 68.7% (standard deviation: 16.4) in HRCT group
- Resting oxyhaemoglobin saturation: 93.2% (standard deviation: 7.8) in the surgical biopsy group, 96.6% (standard deviation: 2.5) in the HRCT group
- 6-minute walk distance: not stated
- Symptom assessment: not assessed
- Use of systemic corticosteroid therapy: 89%
- Use of other therapy: azathioprine 27%, cyclophosphamide 20%, colchicine 20%

**Outcomes**
- Duration of survival

**Notes**
- Funding source: not stated

Antoniou 2008 [26]

**Study Design**
- Type of Study: retrospective cohort
- Trial Design: single centre
- Country/ies: United Kingdom

**Study Duration**
- 3 years

**Participants**
- Number: 249
- Definition of diagnosis: surgical biopsy, clinical and HRCT criteria: (1) bilateral basal or widespread crackles; (2) restrictive ventilatory defect or isolated depression of DLCO; (3) computed tomography (CT) appearances indicative of IPF with predominantly basal and subpleural microcystic or macrocystic honeycombing, with variably extensive ground-glass and reticular abnormalities but no consolidation, nodular abnormalities, or other parenchymal abnormalities (apart from centrilobular emphysema); and (4) no environmental exposure to a fibrogenic agent or connective tissue disease
- Age: mean 62.5 years (standard deviation: 10.3)
- Gender: 74%
- Ethnicity: not stated
- Smoking status: 75% ever smoker
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: 16%
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results (% predicted): mean FEV₁ 70.4 (standard deviation: 20.1); mean FVC 68.2 (standard deviation: 22.6); mean DLCO 36.1 (standard deviation: 15.5)
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: not assessed
| Study Design | Participants | Study Duration | Outcomes | Notes |
|--------------|--------------|----------------|----------|-------|
| Type of Study: retrospective cohort | Number: 99 (86 included) | 1 year | Proportion of mortality | Funding source: not stated |
| Trial Design: single centre | Definition of diagnosis: histological diagnosis, clinical history, and chest X-Ray or computed tomography | | | |
| Country/ies: Japan | Age: mean 79.9 years (standard deviation: 6.1) for male, 81.5 years (standard deviation: 7.2) for female | | | |
| | Gender: 60% male | | | |
| | Ethnicity: not stated | | | |
| | Smoking status: 66% ever smoker | | | |
| | Use of home oxygen therapy: 12% | | | |
| | Time since diagnosis: not stated | | | |
| | Percentage of patients with surgical lung biopsy: not stated | | | |
| | Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated | | | |
| | Lung function results: mean FEV\textsubscript{1} 68.8% predicted (standard deviation: 24.2) for male, 74.8 (27.8) for female; mean FVC 2.0L (standard deviation: 0.72) for male, 1.34 L (0.58) for female; mean DLCO 67% predicted (standard deviation: 31.8) for male, 51 (24.2) for female | | | |
| | Resting oxyhaemoglobin saturation: not stated | | | |
| | 6-minute walk distance: not stated | | | |
| | Symptom assessment: not stated | | | |
| | Use of systemic corticosteroid therapy: not stated | | | |
| | Use of other therapy: not stated | | | |
| Ashley 2016 [28] | Type of Study: prospective cohort | 80 weeks | Duration of survival | Funding source: Not stated |
| Trial Design: national multicentre (n = 9) | | | | |
| Country/ies: United States | | | | |
| | Number: 60 | | | |
| | Definition of diagnosis: characteristic computed tomography findings or usual interstitial pneumonia pathology on biopsy | | | |
| | Age: mean 64.6 years (standard deviation: 7.7) | | | |
| | Gender: 68% male | | | |
| | Ethnicity: not stated | | | |
| | Smoking status: 68% ever smoker | | | |
| | Use of home oxygen therapy: not stated | | | |
| | Time since diagnosis: not stated | | | |
| | Percentage of patients with surgical lung biopsy: not stated | | | |
| | Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated | | | |
| Study Design | Participants | Outcomes | Notes |
|--------------|--------------|----------|-------|
| • Lung function results (% predicted): mean FVC 70 (standard deviation: 16.2); mean DLCO 46.1 (standard deviation: 13.1)  
• Resting oxyhaemoglobin saturation: not stated  
• 6-minute walk distance: not stated  
• Symptom assessment: not assessed  
• Use of systemic corticosteroid therapy: 10%  
• Use of other therapy: mycophenolate mofetil: 2%, azathioprine 5% | • Number: 113  
• Definition of diagnosis: the 2002 American Thoracic Society/European Respiratory Society International Multidisciplinary Consensus Classification of the Idiopathic Interstitial Pneumonias Statement  
• Age: mean 61.9 years (standard deviation: 12.7)  
• Gender: 80% male  
• Ethnicity: not stated  
• Smoking status: 66% ever smoker  
• Use of home oxygen therapy: not stated  
• Time since diagnosis: not stated  
• Percentage of patients with surgical lung biopsy: 72%  
• Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated  
• Lung function results (% predicted): mean FEV1 74.8 (standard deviation: 23.1); FVC 60.9 (standard deviation: 19.6); DLCO 43.6 (standard deviation: 16.5)  
• Resting oxyhaemoglobin saturation: not stated  
• 6-minute walk distance: not stated  
• Symptom assessment: not assessed  
• Use of systemic corticosteroid therapy: 84%  
• Use of other therapy: azathioprine/cyclophosphamide 42%, acetylcysteine 50% | | • Funding source: public  
• Author provided additional data for clarification of study duration and survival/mortality. |

**Barlo 2009 [29]**

**Study Design**
- Type of Study: retrospective cohort
- Trial Design: national multicentre (n = 2)
- Country/ies: Netherlands

**Study Duration**
- At least 1 year

**Participants**
- Number: 113
- Definition of diagnosis: the 2002 American Thoracic Society/European Respiratory Society International Multidisciplinary Consensus Classification of the Idiopathic Interstitial Pneumonias Statement
- Age: mean 61.9 years (standard deviation: 12.7)
- Gender: 80% male
- Ethnicity: not stated
- Smoking status: 66% ever smoker
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: 72%
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results (% predicted): mean FEV1 74.8 (standard deviation: 23.1); FVC 60.9 (standard deviation: 19.6); DLCO 43.6 (standard deviation: 16.5)
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: not assessed
- Use of systemic corticosteroid therapy: 84%
- Use of other therapy: azathioprine/cyclophosphamide 42%, acetylcysteine 50%

**Outcomes**
- Proportion of mortality

**Notes**
- Funding source: unclear

**Bennett 2015 [30]**

**Study Design**
- Type of Study: retrospective cohort
- Trial Design: single centre
- Country/ies: Italy

**Study Duration**
- At least 1 year

**Participants**
- Number: 90
- Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management
- Age: not stated
• Gender: not stated
• Ethnicity: not stated
• Smoking status: not stated
• Use of home oxygen therapy: not stated
• Time since diagnosis: not stated
• Percentage of patients with surgical lung biopsy: not stated
• Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
• Lung function results: not stated
• Resting oxyhaemoglobin saturation: not stated
• 6-minute walk distance: not stated
• Symptom assessment: not stated
• Use of systemic corticosteroid therapy: not stated
• Use of other therapy: not stated

Outcomes

Proportion of mortality

Notes

• Funding source: unclear
• Author provided additional data for clarification of study duration and survival/mortality.

Bhattacharyya 2009 [31]

Study Design

• Type of Study: prospective cohort
• Trial Design: single centre
• Country/ies: India

Study Duration

1 year

Participants

• Number: 7
• Definition of diagnosis: clinical and radiological diagnosis
• Age: mean 72.4 +/- 7 years
• Gender: 71% male
• Ethnicity: not stated
• Smoking status: not stated
• Use of home oxygen therapy: not stated
• Time since diagnosis: not stated
• Percentage of patients with surgical lung biopsy: not stated
• Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
• Lung function results: median 2.04L (range: 0.96-2.89)
• Resting oxyhaemoglobin saturation: median 97% (range: 94-99)
• 6-minute walk distance: not stated
• Symptom assessment: not stated
• Use of systemic corticosteroid therapy: not stated
• Use of other therapy: not stated

Outcomes

Proportion of mortality

Notes

Bjoraker 1998 [32]

Study Design

• Type of Study: retrospective cohort
• Trial Design: single centre
• Country/ies: United States

Study Duration

1 year

Participants

• Number: 104 included
• Definition of diagnosis: Turner-Warwick criteria

Notes

Funding source: not stated
- Age: mean 61.7 years (standard deviation: 10.6)
- Gender: 52% male
- Ethnicity: not stated
- Smoking status: 55% ever smoker
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: 100%
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results (% predicted): mean FEV$_1$ 89 (standard deviation: 21) for UIP group, 86 (22) for others group; mean FVC 79 (standard deviation: 19) for UIP group, 80 (27) for others group; mean DLCO 48 (standard deviation: 13) for UIP group, 50 (16) for others group
- Resting oxyhaemoglobin saturation: mean 91% (standard deviation: 7)
- 6-minute walk distance: not stated
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: 89%
- Use of other therapy: not stated

**Outcomes**
Proportion of mortality

**Notes**
Funding source: not stated

**Bjurstrom 2013 [33]**

| Study Design         |                                      |
|----------------------|--------------------------------------|
| Type of Study:       | retrospective cohort                  |
| Trial Design:        | single centre                         |
| Country/ies:         | Denmark                               |
| **Study Duration**   | 5 years                               |

| Participants         |                                      |
|----------------------|--------------------------------------|
| Number:              | 90                                    |
| Age:                 | 53 +/- 10.2 years                     |
| Gender:              | 68% male                              |
| Ethnicity:           | not stated                            |
| Smoking status:      | not stated                            |
| Use of home oxygen therapy: | not stated                  |
| Time since diagnosis:| not stated                            |
| Percentage of patients with surgical lung biopsy: | not stated                |
| Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: | not stated                |
| Lung function results (% predicted): mean FEV$_1$, 46+/-18, mean FVC 45+/-17%; mean DLCO 24+/-14 |
| Resting oxyhaemoglobin saturation: | not stated                  |
| 6-minute walk distance: | not stated                  |
| Symptom assessment:  | not stated                            |
| Use of systemic corticosteroid therapy: | not stated                  |
| Use of other therapy:| not stated                            |

**Outcomes**
Proportion of mortality

**Notes**
Funding source: not stated

**Boomars 1995 [34]**

| Study Design         |                                      |
|----------------------|--------------------------------------|
| Type of Study:       | retrospective cohort                  |
| Trial Design:        | single centre                         |
| Country/ies:         | Netherlands                           |

134
### Study Duration
At least 2 years

### Participants
- **Number:** 49
- **Definition of diagnosis:** compatible clinical info, evidence of diffuse parenchymal infiltrates on CXR; most patients with restrictive lung function; excluded significant environmental/occupational exposure, extrinsic allergic alveolitis, left ventricular failure, collagen vascular disease
- **Age:** mean 56 years (range: 30-73)
- **Gender:** not stated
- **Ethnicity:** not stated
- **Smoking status:** not stated
- **Use of home oxygen therapy:** not stated
- **Time since diagnosis:** not stated
- **Percentage of patients with surgical lung biopsy:** 100%
- **Percentage of patients with definite usual interstitial pneumonia patterns on HRCT:** not stated
- **Lung function results:** not stated
- **Resting oxyhaemoglobin saturation:** not stated
- **6-minute walk distance:** not stated
- **Symptom assessment:** not stated
- **Use of systemic corticosteroid therapy:** 45%
- **Use of other therapy:** not stated

### Outcomes
- **Proportion of mortality**

### Notes
- **Funding source:** industry

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### Bournazos 2011 [35]

#### Study Design
- **Type of Study:** prospective cohort
- **Trial Design:** single centre
- **Country/ies:** UK

#### Study Duration
1 year

#### Participants
- **Number:** 142
- **Definition of diagnosis:** the 2008 Interstitial lung disease guideline: the British Thoracic Society in collaboration with the Thoracic Society of Australia and New Zealand and the Irish Thoracic Society
- **Age:** mean 70 years (standard deviation: 8.8)
- **Gender:** 66.9% male
- **Ethnicity:** not stated
- **Smoking status:** not stated
- **Use of home oxygen therapy:** not stated
- **Time since diagnosis:** not stated
- **Percentage of patients with surgical lung biopsy:** not stated
- **Percentage of patients with definite usual interstitial pneumonia patterns on HRCT:** not stated
- **Lung function results (% predicted):** mean FEV1 87.52 (standard deviation: 20); mean FVC 87.65 (standard deviation: 19.6); mean DLCO 52.75 (standard deviation: 15.9)
- **Resting oxyhaemoglobin saturation:** not stated
- **6-minute walk distance:** not stated
- **Symptom assessment:** not assessed
- **Use of systemic corticosteroid therapy:** not stated
- **Use of other therapy:** not stated

#### Outcomes
- **Proportion of patients with disease progression**
### Cai 2014 [36]

#### Study Design
- Type of Study: prospective cohort
- Trial Design: single centre
- Country/ies: China

#### Study Duration
- At least 1 year

#### Participants
- Number: 210
- Definition of diagnosis: the 2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment International Consensus Statement
- Age: mean 64 years (standard deviation: 10)
- Gender: 73% male
- Ethnicity: Han Chinese 95%
- Smoking status: 69% ever smoker
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: 5%
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results:
  - Resting oxyhaemoglobin saturation: not stated
  - 6-minute walk distance: not stated
  - Symptom assessment: not assessed
  - Use of systemic corticosteroid therapy: not stated
  - Use of other therapy: immunosuppressants 48%, N-acetylcysteine 5%, interferon-gamma 0.5%

### Castria 2012 [37]

#### Study Design
- Type of Study: retrospective cohort
- Trial Design: single centre
- Country/ies: Italy

#### Study Duration
- 1 year

#### Participants
- Number: 126
- Definition of diagnosis: the 2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment International Consensus Statement
- Age: mean 66.5 years (standard deviation: 2.8)
- Gender: 74% male
- Ethnicity: not stated
- Smoking status: 46.8% ex-smoker
- Use of home oxygen therapy: 52% at rest
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: not stated
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results (% predicted): mean FEV$_1$ 62.6 (standard deviation: 22.4); mean FVC 57.6 (standard deviation: 19.6); mean DLCO 33 (standard deviation: 16)
- Resting oxyhaemoglobin saturation: not stated
### Outcomes
- Proportion of mortality

### Notes
- Funding source: not stated

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**Cayon 2010 [38]**

#### Study Design
- Type of Study: prospective cohort
- Trial Design: national multicentre (n = 2)
- Country/ies: Cuba

#### Study Duration
- 1 year

#### Participants
- Number: 12
- Definition of diagnosis: histologically verified
- Age: median 56 years (interquartile range: 39-66)
- Gender: 50% male
- Ethnicity: Caucasian 58.3%
- Smoking status: 25% ever smoker
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: 8.3%
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results (% predicted): mean FVC 59.3 (standard deviation: 8.4)
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: modified Medical Research Council Dyspnoea score: mean 2.0 (standard deviation 1.0)
- Use of systemic corticosteroid therapy: 58.3%
- Use of other therapy: azathioprine: 16.7%

#### Outcomes
- Proportion of mortality

#### Notes
- Funding source: unclear

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**Civic 2012 [39]**

#### Study Design
- Type of Study: retrospective cohort
- Trial Design: single centre
- Country/ies: United States

#### Study Duration
- At least 2 years

#### Participants
- Number: 43
- Definition of diagnosis: not stated
- Age: mean 65 years (standard deviation: 10)
- Gender: 63% male
- Ethnicity: not stated
- Smoking status: 63% ever smokers
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: not stated
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results (% predicted): mean FVC 70 (standard deviation:
| Study Design | Type of Study: retrospective cohort |
| Study Duration | 1 year |
| Participants | Number: 52 |
| E; | Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management Age: mean 63.2 years (standard deviation 7.9) |
| Gender: 62% male |
| Ethnicity: not stated |
| Smoking status: 63% ever smoker |
| Use of home oxygen therapy: not stated |
| Time since diagnosis: not stated |
| Percentage of patients with surgical lung biopsy: not stated |
| Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated |
| Lung function results (% predicted): mean FVC 72.5 (standard deviation: 16.6); mean DLCO 45.8 (standard deviation: 12) |
| Resting oxyhaemoglobin saturation: not stated |
| 6-minute walk distance: not stated |
| Symptom assessment: not stated |
| Use of systemic corticosteroid therapy: not stated |
| Use of other therapy: not stated |
| Outcomes | Change in forced vital capacity |
| Notes | Funding source: not stated |
| Study Design | • Type of Study: prospective cohort  
|             | • Trial Design: single centre  
|             | • Country/ies: United States |
| Study Duration | 1 year |
| Participants | • Number: 17  
|              | • Definition of diagnosis: the 2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment International Consensus Statement  
|              | • Age: mean 67 years (range: 58-81)  
|              | • Gender: 76% male  
|              | • Ethnicity: not stated  
|              | • Smoking status: not stated  
|              | • Use of home oxygen therapy: 9 (53%)  
|              | • Time since diagnosis: not stated  
|              | • Percentage of patients with surgical lung biopsy: not stated  
|              | • Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated  
|              | • Lung function results (% predicted): mean FEV$_1$ 85.44 (standard deviation: 3.79); mean FVC 73.96 (standard deviation: 3.76); mean DLCO 43.65 (standard deviation: 16.91)  
|              | • Resting oxyhaemoglobin saturation: not stated  
|              | • 6-minute walk distance: mean 464.89m (standard deviation: 249.54)  
|              | • Symptom assessment: not assessed  
|              | • Use of systemic corticosteroid therapy: not stated  
|              | • Use of other therapy: not stated |
| Outcomes | • Proportion of mortality  
| Notes | • Change in forced vital capacity  

Funding source: public
### Treatment International Consensus Statement

- **Age**: mean 66 years (range: 47-79)
- **Gender**: 78% male
- **Ethnicity**: not stated
- **Smoking status**: 72% ever smoker
- **Use of home oxygen therapy**: not stated
- **Time since diagnosis**: not stated
- **Percentage of patients with surgical lung biopsy**: 42%
- **Percentage of patients with definite usual interstitial pneumonia patterns on HRCT**: not stated
- **Lung function results (% predicted)**: mean FVC 64.4; mean DLCO 39.8
- **Resting oxyhaemoglobin saturation**: not stated
- **6-minute walk distance**: mean 415m
- **Symptom assessment**: not stated
- **Use of systemic corticosteroid therapy**: not allowed
- **Use of other therapy**: not allowed

### Outcomes
- Proportion of mortality
- Respiratory-specific mortality
- Change in forced vital capacity
- Change in diffusing capacity for carbon monoxide

### Notes
- Funding source: mixed

### Demedics 2005 [177]

#### Study Design
- **Type of Study**: randomised controlled trial
- **Trial Design**: phase 3, parallel-group, international multicentre (number of centres not stated)
- **Country/ies**: Belgium, France, Germany, Italy, Netherlands, Spain, United Kingdom

#### Study Duration
- 1 year

#### Participants
- **Number**: 75 (placebo group)
- **Definition of diagnosis**: the 2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment International Consensus Statement
- **Age**: mean 64 years (standard deviation: 9)
- **Gender**: 75% male
- **Ethnicity**: not stated
- **Smoking status**: 69.4% ever smoker
- **Use of home oxygen therapy**: not stated
- **Time since diagnosis**: 56% within 6 months
- **Percentage of patients with surgical lung biopsy**: 47%
- **Percentage of patients with definite usual interstitial pneumonia patterns on HRCT**: not stated
- **Lung function results (% predicted)**: mean FVC 66.57 (standard deviation: 14.42); mean DLCO 44.79 (standard deviation: 15.15)
- **Resting oxyhaemoglobin saturation**: not stated
- **6-minute walk distance**: not stated
- **Symptom assessment**: total St George’s Respiratory Questionnaire score: mean 52 (standard deviation: 16); dyspnoea score: mean 7.92 (standard deviation: 3.99)
- **Use of systemic corticosteroid therapy**: 100%
- **Use of other therapy**: azathioprine 100%

#### Outcomes
- Proportion of mortality
| Notes | Proportion of patients with disease progression |
|-------|----------------------------------------------|
| Notes | Funding source: industry                     |

Diaz 2012 [43]

| Study Design | Type of Study: prospective cohort |
|-------------|----------------------------------|
|             | Trial Design: single centre       |
|             | Country/ies: United States        |

| Study Duration | 80-130 weeks |
|----------------|-------------|

| Participants | Number: 10 |
|--------------|------------|
|              | Definition of diagnosis: the 2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment International Consensus Statement |
|              | Age: mean 68.4 years (standard error: 1.82) |
|              | Gender: 80% male |
|              | Ethnicity: not stated |
|              | Smoking status: 50% ever smoker |
|              | Use of home oxygen therapy: none |
|              | Time since diagnosis: not stated |
|              | Percentage of patients with surgical lung biopsy: 40% |
|              | Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated |
|              | Lung function results (% predicted): mean FVC 79.2 (standard error: 4.27); mean DLCO 46.6 (standard error: 3.24) |
|              | Resting oxyhaemoglobin saturation: not stated |
|              | 6-minute walk distance: mean 386m (standard error: 41.35) |
|              | Symptom assessment: not assessed |
|              | Use of systemic corticosteroid therapy: none |
|              | Use of other therapy: none |

| Outcomes | Proportion of mortality |
|---------|-------------------------|
| Notes   | Funding source: mixed   |

Erbes 1997 [44]

| Study Design | Type of Study: retrospective cohort |
|--------------|-----------------------------------|
|              | Trial Design: single centre        |
|              | Country/ies: Germany               |

| Study Duration | At least 1 year |
|----------------|-----------------|

| Participants | Number: 99 |
|--------------|------------|
|              | Definition of diagnosis: histologically confirmed |
|              | Age: mean 53.2 years (standard deviation: 15.4) |
|              | Gender: 52% male |
|              | Ethnicity: not stated |
|              | Smoking status: 56% current smoker |
|              | Use of home oxygen therapy: not stated |
|              | Time since diagnosis: not stated |
|              | Percentage of patients with surgical lung biopsy: 96% |
|              | Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated |
|              | Lung function results (% predicted): mean FEV\textsubscript{1} 69.16 (95% CI: 64.9-73.5); mean FVC 89.19 (95% CI: 83.5-95.1); mean DLCO 46.3 (95% CI: 41.6-51) |
|              | Resting oxyhaemoglobin saturation: not stated |
| Faverio 2015 [45] |  |
| --- | --- |
| **Study Design** | Type of Study: retrospective cohort  
Trial Design: single centre  
Country/ies: Italy |
| **Study Duration** | 2 years |
| **Participants** | Number: 11  
Definition of diagnosis: unclear  
Age: median 68 years  
Gender: 6% male  
Ethnicity: not stated  
Smoking status: not stated  
Use of home oxygen therapy: not stated  
Time since diagnosis: not stated  
Percentage of patients with surgical lung biopsy: not stated  
Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated  
Lung function results: not stated  
Resting oxyhaemoglobin saturation: not stated  
6-minute walk distance: not stated  
Symptom assessment: not assessed  
Use of systemic corticosteroid therapy: 100%  
Use of other therapy: cyclophosphamide 100% |
| **Outcomes** | Proportion of mortality |
| **Notes** | Funding source: unclear |

| Fiorucci 2008 [46] |  |
| --- | --- |
| **Study Design** | Type of Study: prospective cohort  
Trial Design: single centre  
Country/ies: Italy |
| **Study Duration** | 3 years |
| **Participants** | Number: 30  
Definition of diagnosis: the 2002 American Thoracic Society/European Respiratory Society International Multidisciplinary Consensus Classification of the Idiopathic Interstitial Pneumonias Statement  
Age: mean 65 years (standard deviation: 2.3) for group 1, 65.1 (2.7) for group 2, 65.2 (3.2) for group 3  
Gender: 57% male  
Ethnicity: not stated  
Smoking status: not stated  
Use of home oxygen therapy: not stated  
Time since diagnosis: not stated  
Percentage of patients with surgical lung biopsy: not stated  
Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated |
| **Outcomes** |  |
| **Notes** | Funding source: unclear |
## Lung Function Results

- Mean FEV1 (% predicted): 77.6 (standard deviation 19.2) for group 1 (higher lymphocyte count), 71.8 (18.7) for group 2 (lower lymphocyte count); mean FVC 77 (standard deviation 19.2) for group 1, 71 (18.7) for group 2; mean DLCO 59.5 (standard deviation 22.5) for group 1, 51.6 (21.7) for group 2
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: Dyspnoea score: mean 5.7 (standard deviation: 1.3) for group 1, 5.3 (1.3) for group 2, 8.4 (2.5) for group 3
- Use of systemic corticosteroid therapy: 100%
- Use of other therapy: cyclophosphamide 30%, colchicine 33%

## Outcomes

| Study Design | Notes |
|--------------|-------|
| Type of Study: prospective cohort | Proportion of mortality |
| Trial Design: single centre | |
| Country/ies: Israel | |

## Study Duration

1 year

## Participants

- Number: 46
- Definition of diagnosis: either evidence of diffuse parenchymal infiltrates (peripheral and reticular nodular with lower lobe predominance) on chest radiography or restrictive lung function with a lung biopsy demonstrating varying degrees of interstitial fibrosis and intra-alveolar inflammatory cells
- Age: 64 years +/- 12.8 years
- Gender: 50% male
- Ethnicity: not stated
- Smoking status: 28% ever smoker
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: 7%
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results (% predicted): mean FEV1 77.6 (standard deviation 19.2) for group 1 (higher lymphocyte count), 71.8 (18.7) for group 2 (lower lymphocyte count); mean FVC 77 (standard deviation 19.2) for group 1, 71 (18.7) for group 2; mean DLCO 59.5 (standard deviation 22.5) for group 1, 51.6 (21.7) for group 2
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: 77%
- Use of other therapy: not stated

## Outcomes

| Study Design | Notes |
|--------------|-------|
| Type of Study: retrospective cohort | Proportion of mortality |
| Trial Design: single centre | |
| Country/ies: Canada | |

## Study Duration

1 year

## Participants

- Number: 302
- Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis

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**Fireman 1998 [47]**

| Study Design | Notes |
|--------------|-------|
| Type of Study: prospective cohort | Proportion of mortality |
| Trial Design: single centre | |
| Country/ies: Israel | |

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**Fisher 2017a [48]**

| Study Design | Notes |
|--------------|-------|
| Type of Study: retrospective cohort | Proportion of mortality |
| Trial Design: single centre | |
| Country/ies: Canada | |

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**Funding source**: unclear
### Fisher 2017b [49]

#### Study Design
- Type of Study: retrospective cohort
- Trial Design: single centre
- Country/ies: United States

#### Study Duration
- At least 1 year

#### Participants
- Number: 286
- Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management
- Age: median 67 years (range 43-80)
- Gender: 81.8% male
- Ethnicity: Caucasian 94.6%
- Smoking status: not stated
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: not stated
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results (% predicted): median FVC 68.0 (range 50-114); median DLCO 46.0 (range 30-102)
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: not stated
- Use of other therapy: not stated

#### Outcomes
- Proportion of mortality

#### Notes
- Funding source: public

### Fujimoto 2012 [50]

#### Study Design
- Type of Study: retrospective cohort
**Trial Design:** single centre  
**Country/ies:** Japan  

**Study Duration**  
1 year  

**Participants**  
- Number: 60  
- Definition of diagnosis: the 2002 American Thoracic Society/European Respiratory Society International Multidisciplinary Consensus Classification of the Idiopathic Interstitial Pneumonias Statement  
- Age: median 71 years (range 37-87)  
- Gender: 82% male  
- Ethnicity: not stated  
- Smoking status: 80% ever smokers  
- Use of home oxygen therapy: not stated  
- Time since diagnosis: median 17 months (range 1-120)  
- Percentage of patients with surgical lung biopsy: not stated  
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated  
- Lung function results: not stated  
- Resting oxyhaemoglobin saturation: not stated  
- 6-minute walk distance: not stated  
- Symptom assessment: not stated  
- Use of systemic corticosteroid therapy: not stated  
- Use of other therapy: not stated  

**Outcomes**  
Proportion of mortality  

**Notes**  
- Funding source: public  
- Author provided additional data for clarification of study duration and survival/mortality.  

**Gay 1998 [51]**  

**Study Design**  
- Type of Study: prospective cohort  
- Trial Design: single centre  
- Country/ies: United States  

**Study Duration**  
1 year  

**Participants**  
- Number: 38  
- Definition of diagnosis: symptoms, physiologic abnormalities, or radiographic findings with open lung biopsy confirmation  
- Age: mean 54.6 +/- 2.2 years  
- Gender: 45% male  
- Ethnicity: not stated  
- Smoking status: 71% ever smoker  
- Use of home oxygen therapy: not stated  
- Time since diagnosis: not stated  
- Percentage of patients with surgical lung biopsy: not stated  
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated  
- Lung function results (% predicted): mean FEV1 78.3 (standard deviation 2.9); mean FVC 69.7 (standard deviation 2.5); mean DLCO 49.9 (standard deviation: 2.4)  
- Resting oxyhaemoglobin saturation: note stated  
- 6-minute walk distance: not stated  
- Symptom assessment: not stated  
- Use of systemic corticosteroid therapy: 100%  
- Use of other therapy: not stated
| **Outcomes** | Proportion of mortality |
|-------------|-------------------------|
| **Notes**   | Funding source: public  |

**Gu 2014 [52]**

**Study Design**
- Type of Study: retrospective cohort
- Trial Design: national multicentre (n = 2)
- Country/ies: China

**Study Duration**
- At least 1 year

**Participants**
- Number: 25
- Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management
  - Age: mean 57.3 years (standard deviation 10.7)
  - Gender: 68% male
  - Ethnicity: not stated
  - Smoking status: 40% ever smokers
  - Use of home oxygen therapy: not stated
  - Time since diagnosis: 87 months (standard deviation 40)
  - Percentage of patients with surgical lung biopsy: 100%
  - Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
  - Lung function results: not stated
  - Resting oxyhaemoglobin saturation: not stated
  - 6-minute walk distance: not stated
  - Symptom assessment: not stated
  - Use of systemic corticosteroid therapy: not stated
  - Use of other therapy: not stated

**Outcomes**

**Notes**

| **Outcomes** | Proportion of mortality |
|-------------|-------------------------|
| **Notes**   | Funding source: unclear |

**Hallstrand 2005 [53]**

**Study Design**
- Type of Study: prospective cohort
- Trial Design: single centre
- Country/ies: United States

**Study Duration**
- At least 1 year

**Participants**
- Number: 28
- Definition of diagnosis: the 2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment International Consensus Statement
  - Age: mean 62.7 years (interquartile range 57-69)
  - Gender: 67.9% male
  - Ethnicity: Caucasian 96.4%, other 3.6%
  - Smoking status: 67.9% ever smoker
  - Use of home oxygen therapy: not stated
  - Time since diagnosis: mean 3.1 years (interquartile range 0.8-4.0)
  - Percentage of patients with surgical lung biopsy: 50%
  - Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
  - Lung function results (% predicted): mean FEV₁ 61.1 (interquartile range 45.3-70.8); mean FVC 59.9 (interquartile range 42.4-71.5); mean DLCO 33.0 (interquartile range 23-43.3)
  - Resting oxyhaemoglobin saturation: not stated
### Hamada 2007 [54]

**Study Design**
- Type of Study: prospective cohort
- Trial Design: single centre
- Country/ies: Japan

**Study Duration**
- 5 years

**Participants**
- Number: 61
- Definition of diagnosis: the 2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment International Consensus Statement
- Age: mean 62 years (standard deviation 8)
- Gender: 86% male
- Ethnicity: not stated
- Smoking status: 79% ever smoker
- Use of home oxygen therapy: 27.9%
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: 77%
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results (% predicted): mean FVC 76 (standard deviation 22); mean DLCO 45 (standard deviation 15)
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: 42.6%
- Use of other therapy: not stated

**Outcomes**
- Proportion of mortality
- Duration of survival

**Notes**
- Funding source: unclear

### Hanson 1995 [55]

**Study Design**
- Type of Study: retrospective cohort
- Trial Design: single centre
- Country/ies: United States

**Study Duration**
- At least 1 year

**Participants**
- Number: 58
- Definition of diagnosis: Three criteria: CXR with diffuse reticulation, lung biopsy with interstitial fibrosis, and no other known causes of pulmonary fibrosis
- Age: mean 59.9 years (range 27-84)
- Gender: 47% male
- Ethnicity: not stated
- Smoking status: not stated
- Use of home oxygen therapy: not stated
### Outcomes
- Proportion of mortality
- Proportion of patients with disease progression
- Duration of survival

### Notes
- Funding source: unclear

#### Harris 2010 [56]

**Study Design**
- Type of Study: prospective cohort
- Trial Design: national (the British Thoracic Society cryptogenic fibrosing alveolitis study)
- Country/ies: United Kingdom

**Study Duration**
- 1 year

**Participants**
- Number: 588
- Definition of diagnosis: codes using the International Classification of Diseases, ninth revision (ICD-9)
- Age: not stated
- Gender: not stated
- Ethnicity: not stated
- Smoking status: not stated
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: not stated
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results: mean FVC 2.27L (range 0.98 to 4.04); mean DLCO 9.22 (range 2.53-23.5)
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: not stated
- Use of other therapy: not stated

**Outcomes**
- Proportion of mortality

**Notes**
- Funding source: public

#### Hiwatari 1997 [57]

**Study Design**
- Type of Study: retrospective cohort
- Trial Design: single centre
- Country/ies: Japan

**Study Duration**
- At least 1 year

**Participants**
- Number: 48
- Definition of diagnosis: a combination of medical records, clinical and
| Study Design | Holland 2013 [58] |
|--------------|-------------------|
| **Type of Study:** | prospective cohort |
| **Trial Design:** | national multicentre (n = 2) |
| **Country/ies:** | Australia |
| **Study Duration** | 4 years |
| **Participants** | |
| Number: | 40 |
| Definition of diagnosis: | the 2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment International Consensus Statement |
| Age: | mean 73 years (standard deviation 7) |
| Gender: | not stated |
| Ethnicity: | not stated |
| Smoking status: | not stated |
| Use of home oxygen therapy: | not stated |
| Time since diagnosis: | not stated |
| Percentage of patients with surgical lung biopsy: | not stated |
| Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: | not stated |
| Lung function results (% predicted): | mean FVC 74 (standard deviation 20); mean DLCO 46 (standard deviation 17) |
| Resting oxyhaemoglobin saturation: | not stated |
| 6-minute walk distance: | mean 350m (standard deviation 124) |
| Symptom assessment: | not stated |
| Use of systemic corticosteroid therapy: | not stated |
| Use of other therapy: | not stated |
| **Outcomes** | Proportion of mortality |
| **Notes** | Funding source: public |

Holland 2013 [58]
| Study Design       | • Type of Study: retrospective cohort  
|                   | • Trial Design: national using the Discharge Abstract Database and the National Ambulatory Care Reporting System from the Canadian Institute for Health Information  
|                   | • Country/ies: Canada  
| Study Duration    | At least 1 year  
| Participants      | • Number: unclear  
|                   | • Definition of diagnosis: codes using the International Classification of Diseases, Version 10, Canadian (ICD-10 CA)  
|                   | • Age: not stated  
|                   | • Gender: not stated  
|                   | • Ethnicity: not stated  
|                   | • Smoking status: not stated  
|                   | • Use of home oxygen therapy: not stated  
|                   | • Time since diagnosis: not stated  
|                   | • Percentage of patients with surgical lung biopsy: not stated  
|                   | • Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated  
|                   | • Lung function results: not stated  
|                   | • Resting oxyhaemoglobin saturation: not stated  
|                   | • 6-minute walk distance: not stated  
|                   | • Symptom assessment: not stated  
|                   | • Use of systemic corticosteroid therapy: not stated  
|                   | • Use of other therapy: not stated  
| Outcomes          | Proportion of mortality  
| Notes             | Funding source: industry  

Hosein 2016 [60]

| Study Design       | • Type of Study: retrospective cohort  
|                   | • Trial Design: single centre  
|                   | • Country/ies: Canada  
| Study Duration    | 1 year  
| Participants      | • Number: 42 (22 untreated)  
|                   | • Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management  
|                   | • Age: mean 68 years (standard deviation 9)  
|                   | • Gender: 68% male  
|                   | • Ethnicity: not stated  
|                   | • Smoking status: not stated  
|                   | • Use of home oxygen therapy: not stated  
|                   | • Time since diagnosis: not stated  
|                   | • Percentage of patients with surgical lung biopsy: 32%  
|                   | • Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated  
|                   | • Lung function results (% predicted): mean FEV$_1$ 77 (standard deviation 17); mean FVC 76 (standard deviation 18); mean DLCO 40 (standard deviation 12)  
|                   | • Resting oxyhaemoglobin saturation: not stated  
|                   | • 6-minute walk distance: mean 396m (standard deviation 123)  
|                   | • Symptom assessment: modified Medical Research Council Dyspnoea score: mean 2.4 (standard deviation 0.8)  

150
| Study Design | Hubbard 1998 [61] |
|--------------|-------------------|
| **Type of Study:** | prospective cohort |
| **Trial Design:** | national multicentre (n = 9) |
| **Country/ies:** | United Kingdom |
| **Study Duration:** | At least 18 months |
| **Participants** |  |
| **Number:** | 244 (76 incident cases) |
| **Definition of diagnosis:** | histological confirmation or basal inspiratory crackles plus no documented history of exposure to asbestos |
| **Age:** | 69.7 years (standard deviation 9.6) |
| **Gender:** | Male:Female ratio 2.8:1 |
| **Ethnicity:** | not stated |
| **Smoking status:** | not stated |
| **Use of home oxygen therapy:** | not stated |
| **Time since diagnosis:** | not stated |
| **Percentage of patients with surgical lung biopsy:** | not stated |
| **Percentage of patients with definite usual interstitial pneumonia patterns on HRCT:** | not stated |
| **Lung function results (% predicted):** | mean FVC 78.5 (standard error 2.8), mean DLCO 49.2 (standard error 2.2) |
| **Resting oxyhaemoglobin saturation:** | not stated |
| **6-minute walk distance:** | not stated |
| **Symptom assessment:** | not stated |
| **Use of systemic corticosteroid therapy:** | 47% |
| **Use of other therapy:** | cyclophosphamide 6.8%, azathioprine 2.6% |
| **Outcomes** | Proportion of mortality |
| **Notes** | Funding source: unclear |

| Study Design | Huynh 2015 [62] |
|--------------|-----------------|
| **Type of Study:** | retrospective cohort |
| **Trial Design:** | single centre |
| **Country/ies:** | United States |
| **Study Duration:** | 1 year |
| **Participants** |  |
| **Number:** | 70 |
| **Definition of diagnosis:** | not stated |
| **Age:** | not stated |
| **Gender:** | not stated |
| **Ethnicity:** | not stated |
| **Smoking status:** | not stated |
| **Use of home oxygen therapy:** | not stated |
| **Time since diagnosis:** | not stated |
| **Percentage of patients with surgical lung biopsy:** | not stated |
| **Percentage of patients with definite usual interstitial pneumonia patterns on HRCT:** | not stated |
| **Lung function results:** | not stated |
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: not stated
- Use of other therapy: not stated

**Outcomes**
Proportion of patients with disease progression

**Notes**
Funding source: unclear

### Inase 2003 [63]

#### Study Design
- Type of Study: retrospective cohort
- Trial Design: single centre
- Country/ies: Japan

#### Study Duration
1 year

#### Participants
- Number: 13
- Definition of diagnosis: surgical lung biopsy or clinical diagnosis with characteristic findings on computed tomography, clubbed fingers, and restrictive lung pattern
- Age: mean 67.1 years (standard deviation 9)
- Gender: 77% male
- Ethnicity: not stated
- Smoking status: 46% ever smoker, 23% unknown
- Use of home oxygen therapy: not stated
- Time since diagnosis: median 0 year (range 0-7)
- Percentage of patients with surgical lung biopsy: not stated
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results: not stated
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: 100%
- Use of other therapy: not stated

#### Outcomes
Proportion of mortality

**Notes**
Funding source: unclear

### Iwasawa 2014 [64]

#### Study Design
- Type of Study: retrospective cohort
- Trial Design: single centre
- Country/ies: Japan

#### Study Duration
1 year

#### Participants
- Number: 40
- Definition of diagnosis: the 2002 American Thoracic Society/European Respiratory Society International Multidisciplinary Consensus Classification of the Idiopathic Interstitial Pneumonias Statement
- Age: mean 68.2 years (standard deviation 8.3)
- Gender: 73% male
- Ethnicity: not stated
- Smoking status: not stated
- Use of home oxygen therapy: 15%
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: not stated
### Study Design
- **Type of Study:** retrospective cohort
- **Trial Design:** national multicentre (n = 48)
- **Country/ies:** Japan

### Study Duration
- 10 years

### Participants
- **Number:** 222
- **Definition of diagnosis:** Open lung biopsy/autopsy
- **Age:** not stated
- **Gender:** 73% male
- **Ethnicity:** not stated
- **Smoking status:** not stated
- **Use of home oxygen therapy:** not stated
- **Time since diagnosis:** not stated
- **Percentage of patients with surgical lung biopsy:** not stated
- **Percentage of patients with definite usual interstitial pneumonia patterns on HRCT:** not stated
- **Lung function results:** not stated
- **Resting oxyhaemoglobin saturation:** not stated
- **6-minute walk distance:** not stated
- **Symptom assessment:** not stated
- **Use of systemic corticosteroid therapy:** not stated
- **Use of other therapy:** not stated

### Outcomes
- Proportion of mortality
- Change in forced vital capacity
- Proportion of patients with disease progression

### Notes
- Funding source: public

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### Izumi 1992 [65]

| Study Design   | Details |
|----------------|---------|
| **Type of Study:** | retrospective cohort |
| **Trial Design:** | national multicentre (n = 48) |
| **Country/ies:** | Japan |

| Study Duration | 10 years |

| Participants   | Details |
|----------------|---------|
| **Number:** | 222 |
| **Definition of diagnosis:** | Open lung biopsy/autopsy |
| **Age:** | not stated |
| **Gender:** | 73% male |
| **Ethnicity:** | not stated |
| **Smoking status:** | not stated |
| **Use of home oxygen therapy:** | not stated |
| **Time since diagnosis:** | not stated |
| **Percentage of patients with surgical lung biopsy:** | not stated |
| **Percentage of patients with definite usual interstitial pneumonia patterns on HRCT:** | not stated |
| **Lung function results:** | not stated |
| **Resting oxyhaemoglobin saturation:** | not stated |
| **6-minute walk distance:** | not stated |
| **Symptom assessment:** | not stated |
| **Use of systemic corticosteroid therapy:** | not stated |
| **Use of other therapy:** | not stated |

| Outcomes | Details |
|----------|---------|
| Proportion of mortality |

| Notes | Details |
|-------|---------|
| Funding source: public |

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### Jacob 2016 [67]

| Study Design   | Details |
|----------------|---------|
| **Type of Study:** | retrospective cohort |
| **Trial Design:** | single centre |
| **Country/ies:** | United Kingdom |

| Study Duration | 1 year |

| Participants   | Details |
|----------------|---------|
| **Number:** | 283 |
| **Definition of diagnosis:** | the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management |
| **Age:** | median 67 years |
| **Gender:** | 77% male |
| **Ethnicity:** | not stated |

| Outcomes | Details |
|----------|---------|
| Proportion of mortality |

| Notes | Details |
|-------|---------|
| Funding source: unclear |

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153
| Study Design | • Smoking status: 66% ever smoker  
• Use of home oxygen therapy: not stated  
• Time since diagnosis: not stated  
• Percentage of patients with surgical lung biopsy: 21%  
• Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated  
• Lung function results (% predicted): mean FEV$_1$ 70.8 (standard deviation 19.1); mean FVC 68.8 (standard deviation 20.5); mean DLCO 36.1 (standard deviation 12.9)  
• Resting oxyhaemoglobin saturation: not stated  
• 6-minute walk distance: not stated  
• Symptom assessment: not stated  
• Use of systemic corticosteroid therapy: not stated  
• Use of other therapy: not stated  

| Outcomes | Duration of survival (Kaplan-Meier Curve – data not extractable)  

| Notes | • Funding source: unclear  
• Author provided additional data for clarification of study duration and survival/mortality.

### Jaffar 2014 [67]

#### Study Design
- Type of Study: prospective cohort  
- Trial Design: international multicentre (n = 3)  
- Country/ies: Australia, USA, Italy

#### Study Duration
- 1 year

#### Participants
- Number: 72  
- Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management  
- Age: mean 68 years (standard deviation 9)  
- Gender: 57% male  
- Ethnicity: not stated  
- Smoking status: 63% ever smoker  
- Use of home oxygen therapy: not stated  
- Time since diagnosis: not stated  
- Percentage of patients with surgical lung biopsy: not stated  
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated  
- Lung function results (% predicted): mean FEV$_1$ 79 (standard deviation 20); mean FVC 74 (standard deviation 20); mean DLCO 41 (standard deviation 16)  
- Resting oxyhaemoglobin saturation: not stated  
- 6-minute walk distance: not stated  
- Symptom assessment: not stated  
- Use of systemic corticosteroid therapy: not stated  
- Use of other therapy: not stated

#### Outcomes
- Proportion of mortality

#### Notes
- Funding source: public  
- Author provided additional data for clarification of study duration and survival/mortality.

### Jeon 2006 [68]

#### Study Design
- Type of Study: retrospective cohort
| Study Design | Type of Study: prospective cohort |
|--------------|----------------------------------|
|              | Trial Design: single centre       |
|              | Country/ies: Czech Republic       |
| Study Duration | At least 1 year, mean 5 years   |
| Participants  | Number: 50                        |
|              | Definition of diagnosis: histologic assessment of lung biopsy or autopsy |
|              | Age: not stated                   |
|              | Gender: 44% male                  |
|              | Ethnicity: not stated              |
|              | Smoking status: not stated         |
|              | Use of home oxygen therapy: not stated |
|              | Time since diagnosis: not stated   |
|              | Percentage of patients with surgical lung biopsy: not stated |
|              | Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated |
|              | Lung function results: not stated  |
|              | Resting oxyhaemoglobin saturation: not stated |
|              | 6-minute walk distance: not stated  |
|              | Symptom assessment: not stated     |
|              | Use of systemic corticosteroid therapy: not stated |
|              | Use of other therapy: not stated   |
| Outcomes     | Proportion of mortality            |
| Notes        | Funding source: unclear            |

Jezek 1979 [69]
### Jezek 1980 [70]

**Study Design**
- Type of Study: prospective cohort
- Trial Design: single centre
- Country/ies: Chechoslovakia

**Study Duration**
6 years

**Participants**
- Number: 56
- Definition of diagnosis: Clinical examination, confirmed by lung biopsy/autopsy/both
- Age: mean 38.6 years (standard deviation 11.6)
- Gender: 43% male
- Ethnicity: not stated
- Smoking status: not stated
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: not stated
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results (% predicted): mean FVC 63.1 (standard deviation 23.4); mean DLCO 35.2 (standard deviation 11.8)
- Resting oxyhaemoglobin saturation: mean 91.5% (standard deviation 5.6)
- 6-minute walk distance: not stated
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: 100%
- Use of other therapy: not stated

**Outcomes**
Proportion of mortality

**Notes**
Funding source: unclear

### Jezkova 1981 [71]

**Study Design**
- Type of Study: retrospective cohort
- Trial Design: single centre
- Country/ies: Czech Republic

**Study Duration**
At least 1 year, mean 5.6 years (standard deviation 3.3)

**Participants**
- Number: 77
- Definition of diagnosis: not stated
- Age: average not stated
- Gender: not stated
- Ethnicity: not stated
- Smoking status: not stated
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: not stated
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results: not stated
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: not stated
- Use of other therapy: not stated

**Notes**
Funding source: unclear
| Outcomes          | Duration of survival |
|-------------------|----------------------|
| Notes             | Funding source: unclear |

**Jo 2017 [72]**

| Study Design                                      |           |
|--------------------------------------------------|-----------|
| Type of Study: prospective cohort                 |           |
| Trial Design: national using the Australian Idiopathic Pulmonary Fibrosis Registry |           |
| Country/ies: Australia                           |           |

| Study Duration | At least 1 year |
|----------------|-----------------|
| Participants   |                 |
| Number: 647 (460 not on antifibrotic therapies) |           |
| Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management |           |
| Age: mean 70.9 years (standard deviation 8.5) |           |
| Gender: 67.7% male |           |
| Ethnicity: not stated |           |
| Smoking status: 71.7% ever smoker |           |
| Use of home oxygen therapy: not stated |           |
| Time since diagnosis: not stated |           |
| Percentage of patients with surgical lung biopsy: not stated |           |
| Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated |           |
| Lung function results (% predicted): mean FVC 81 (standard deviation 21.7); mean 48.4 (standard deviation 16.7) |           |
| Resting oxyhaemoglobin saturation: not stated |           |
| 6-minute walk distance: mean 420m (standard deviation 129) |           |
| Symptom assessment: not stated |           |
| Use of systemic corticosteroid therapy: not stated |           |
| Use of other therapy: not stated |           |

| Outcomes        | Duration of survival |
|-----------------|----------------------|
| Notes           | Funding source: public |
| Author provided additional data for clarification of study duration and survival/mortality. |           |

**Justet 2017 [73]**

| Study Design                                      |           |
|--------------------------------------------------|-----------|
| Type of Study: prospective cohort                 |           |
| Trial Design: single centre                       |           |
| Country/ies: France                              |           |

| Study Duration | 1 year |
|----------------|--------|
| Participants   |        |
| Number: 27     |        |
| Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management |        |
| Age: mean 65.3 years (standard deviation 12.4) |        |
| Gender: 81% male |        |
| Ethnicity: not stated |        |
| Smoking status: 59% ever smoker |        |
| Use of home oxygen therapy: not stated |        |
| Time since diagnosis: mean 2.9 years (standard deviation 2.7) |        |
| Percentage of patients with surgical lung biopsy: not stated |        |
| Percentage of patients with definite usual interstitial pneumonia |        |
| Study Design                  |                                                                                   |
|------------------------------|-----------------------------------------------------------------------------------|
| **Outcomes**                 | • Duration of survival                                                             |
|                              | • Change in forced vital capacity                                                 |
|                              |                                                                                   |
| **Notes**                    | Funding source: unclear                                                             |

Kanematsu 1994 [74]

**Study Design**
- Type of Study: prospective cohort
- Trial Design: single centre
- Country/ies: Japan

**Study Duration**
- 2 years

**Participants**
- Number: 52
- Definition of diagnosis: clinical findings of IPF with pathological findings of usual interstitial pneumonia on open lung biopsy
- Age: mean 58.4 years (standard deviation 6.8) for those without clubbing, 57.3 years (7.9) for those with clubbing
- Gender: 81% male
- Ethnicity: not stated
- Smoking status: 79% ever smoker
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: 100%
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results (% predicted): FVC 78 +/- 21.6 for those without clubbing, 86.9 +/- 24 for those with clubbing; DLCO 51.8 +/- 16 for those without clubbing, 52 +/- 18 for those with clubbing
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: not stated
- Use of other therapy: not stated

**Outcomes**
- Proportion of mortality

**Notes**
- Funding source: public

Khadadah 2003 [75]

**Study Design**
- Type of Study: prospective cohort
- Trial Design: national multicentre (n =2)
- Country/ies: Kuwait

**Study Duration**
- 1 year

**Participants**
- Number: 52
- Definition of diagnosis: the 2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment International Consensus Statement
- Age: mean 55.4 years (standard deviation11.77)
- Gender: 62% male
- Ethnicity: not stated
- Smoking status: 42% ever smokers
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: 56%
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results (% predicted): mean FVC 57.6 (standard deviation 16.78); mean DLCO 55
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: 90%
- Use of other therapy: azathioprine 17%

| Outcomes       | Proportion of mortality |
|----------------|-------------------------|
| Notes          | Funding source: unclear |

**Kim 2012 [76]**

| Study Design       | Type of Study: retrospective cohort |
|--------------------|------------------------------------|
|                    | Trial Design: single centre        |
|                    | Country/ies: Korea                 |

| Study Duration     | At least 16 months                 |
|--------------------|-----------------------------------|

| Participants       | Number: 67 IPF                     |
|--------------------|-----------------------------------|
|                    | Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management |
|                    | Age: mean 69.9 years (standard deviation 9.9) |
|                    | Gender: 64% male                   |
|                    | Ethnicity: not stated              |
|                    | Smoking status: not stated         |
|                    | Use of home oxygen therapy: not stated |
|                    | Time since diagnosis: not stated   |
|                    | Percentage of patients with surgical lung biopsy: not stated |
|                    | Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated |
|                    | Lung function results (% predicted): mean FVC 71.2 (standard deviation 20.2); mean DLCO 67.5 (standard deviation 27) |
|                    | Resting oxyhaemoglobin saturation: not stated |
|                    | 6-minute walk distance: not stated |
|                    | Symptom assessment: not stated     |
|                    | Use of systemic corticosteroid therapy: 23% |
|                    | Use of other therapy: not stated   |

| Outcomes       | Duration of survival |
|----------------|----------------------|
| Notes          | Funding source: unclear |

**Kim 2013 [77]**

| Study Design       | Type of Study: retrospective cohort |
|--------------------|------------------------------------|
|                    | Trial Design: single centre        |
|                    | Country/ies: United States         |

| Study Duration     | More than 5 years                 |
|--------------------|-----------------------------------|

| Participants       | Number: 93                        |
|--------------------|-----------------------------------|

Notes: Funding source: unclear
- Definition of diagnosis: unclear
- Age: mean 64.7 years for rapid group, 62.5 for usual group, 61.7 for long-term group
- Gender: 69% male
- Ethnicity: not stated
- Smoking status: 69.2% ever smoker for rapid group, 78.9% for usual group, 55.6% long-term group
- Use of home oxygen therapy: 83.3% for rapid group, 75.7% for usual group, 73% long-term group
- Time since diagnosis: mean 1.63 years for rapid group, 1.72 for usual group, < 1 for long-term group
- Percentage of patients with surgical lung biopsy: not stated
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: 45.8% for rapid group, 51.4% for usual group, 78.3% for long-term group
- Lung function results (% predicted): mean FVC 62.6 for rapid group, 66.5 for usual group, and 75.3 for long-term group; mean DLCO 40.58 for rapid group, 49.15 for usual group, 51.96 for long-term group
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: mean 296.88m for rapid group, 374.60m for usual group, 426.72m for long-term group
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: 26.9% for rapid group, 27% for usual group, 33.3% for long-term group
- Use of other therapy: N-acetylcysteine: 42.3% for rapid group, 74.4% for usual group, 29.6% for long term group

**Outcomes**
- Proportion of mortality

**Notes**
- Funding source: unclear

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**Kim 2015 [78]**

**Study Design**
- Type of Study: retrospective cohort
- Trial Design: single centre
- Country/ies: Korea

**Study Duration**
- 1 year

**Participants**
- Number: 268
- Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management
- Age: mean 65.9 years (standard deviation 9.6)
- Gender: 67.5% male
- Ethnicity: not stated
- Smoking status: 56.3% ever smoker
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: 20.1%
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results (% predicted): mean FEV₁ 77.8% (standard deviation 18.8); mean FVC: 89.8 (standard deviation 21.5); mean DLCO 65.9 (standard deviation 21.7)
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
| Study Design | Outcomes | Notes |
|--------------|----------|-------|
| • Type of Study: randomised controlled trial | • Proportion of mortality | Funding source: unclear |
| • Trial Design: phase 2, parallel group, international multicentre (n = 29) | • Duration of survival | |
| • Country/ies: Canada, France, Germany, Israel, Italy, Switzerland, United Kingdom, United States | | |
| **Study Duration** | **Participants** | |
| 1 year | • Number: 83 (placebo group) | |
| | • Definition of diagnosis: the 2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment International Consensus Statement | |
| | • Age: mean 65.1 years (standard deviation 9.1) | |
| | • Gender: 76% male | |
| | • Smoking status: current smoker 1.2% | |
| | • Use of home oxygen therapy: 15.5% | |
| | • Time since diagnosis: mean 1.1 years ± SD 1.0 | |
| | • Percentage of patients with surgical lung biopsy: 60.2% | |
| | • Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated | |
| | • Lung function results (% predicted): mean FEV₁ 81.8 (standard deviation 13.8); mean FVC 69.5 (standard deviation 12.6); mean DLCO: 41.4 (standard deviation 9.5) | |
| | • Resting oxyhaemoglobin saturation: mean 96.8% (standard deviation 2.5) | |
| | • 6-minute walk distance: mean 372m (standard deviation 74) | |
| | • Symptom assessment: not stated | |
| | • Use of systemic corticosteroid therapy: 13.1% | |
| | • Use of other therapy: not stated | |
| | **Outcomes** | |
| | • Proportion of mortality | |
| | • Change in 6-minute walk distance | |
| **Notes** | Funding source: industry | |

King 2009 [179]

| Study Design | Outcomes | Notes |
|--------------|----------|-------|
| • Type of Study: randomised controlled trial | • Proportion of mortality | |
| • Trial Design: phase 3, parallel-group, international multicentre (n = 81) | • Duration of survival | |
| • Country/ies: Belgium, Canada, France, Germany, Ireland, Italy, Spain, United Kingdom, United States | | |
| **Study Duration** | **Participants** | |
| Greater than 52 weeks, median 64 weeks | • Number: 275 (placebo group) | |
| | • Definition of diagnosis: the 2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment International Consensus Statement | |
| | • Age: mean 65.9 years (standard deviation 7.9) | |
| | • Gender: 32% male | |
- Ethnicity: Caucasian 95%
- Smoking status: 69% ever smoker
- Use of home oxygen therapy: 15%
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: 55%
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: 85%
- Lung function results (% predicted): mean FVC 73.1 (standard deviation 13.4); mean DLCO 47.3 (standard deviation 9.3)
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: mean 392.8m (standard deviation 112.9)
- Symptom assessment: total St George’s Respiratory Questionnaire score: mean 42.4 (standard deviation 18.2); University of California San Diego Shortness of Breath Questionnaire score: mean 35 (standard deviation 22.7)
- Use of systemic corticosteroid therapy: 17%
- Use of other therapy: not stated

### Outcomes
- Proportion of mortality
- Respiratory-specific mortality
- Change in forced vital capacity
- Change in dyspnoea (University of California, San Diego Shortness of Breath Questionnaire)
- Change in health-related quality of life (St George’s Respiratory Questionnaire)
- Change in 6-minute walk distance

### Notes
- Funding source: industry

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**King 2011 [180]**

### Study Design
- Type of Study: randomised controlled trial
- Trial Design: phase 3, parallel-group, international multicentre (n = 119)
- Country/ies: Australia, Austria, Belgium, Canada, Croatia, Czech Republic, France, Germany, Ireland, Israel, Italy, Japan, Korea, Netherlands, Serbia, Spain, Switzerland, United Kingdom, United States

### Study Duration
- 1 year

### Participants
- Number: 209 (placebo group)
- Definition of diagnosis: the 2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment International Consensus Statement
- Age: mean 63.2 years (standard deviation 9.1)
- Gender: 63.6% male
- Ethnicity: not stated
- Smoking status: 68% ever smoker
- Use of home oxygen therapy: 11%
- Time since diagnosis: median 0.5 years (range 0.05-4.72)
- Percentage of patients with surgical lung biopsy: not stated
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: 46.9%
- Lung function results (% predicted): mean FVC 73.1(standard deviation 15.3); mean DLCO 47.9 (standard deviation 12.7)
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: Dyspnoea Index: mean 7.6 (standard deviation
| Study Design | Participants | Outcomes | Notes |
|--------------|--------------|----------|-------|
| • Type of Study: randomised controlled trial | • Number: 277 (placebo group) | • Proportion of mortality | Funding source: industry |
| • Trial Design: phase 3, parallel-group, international multicentre (n = 129) | • Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management | • Change in forced vital capacity | |
| • Country/ies: Australia, Brazil, Croatia, Israel, Mexico, New Zealand, Peru, Singapore, United States | • Age: mean 67.8 years (standard deviation 7.3) | • Change in diffusing capacity for carbon monoxide | |
| Study Duration | Participants | Outcomes | Notes |
| 1 year | • Ethnicity: Caucasian 90.6% | • Change in dyspnoea (Transition Dyspnoea Index) | |
| • Smoking status: 61% ex-smoker | | | |
| | • Use of home oxygen therapy: 27.4% | | |
| | • Time since diagnosis: mean 1.7 years (standard deviation 1.1) | | |
| | • Percentage of patients with surgical lung biopsy: 28.5% | | |
| | • Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: 94.6% | | |
| | • Lung function results (% predicted): mean FVC 68.6 (standard deviation 10.9); mean DLCO 44.2 (standard deviation 12.5) | | |
| | • Resting oxyhaemoglobin saturation: not stated | | |
| | • 6-minute walk distance: mean 420.7m (standard deviation 98.1) | | |
| | • Symptom assessment: University of California San Diego Shortness of Breath Questionnaire score: mean 36.6 (standard deviation 21.7) | | |
| | • Use of systemic corticosteroid therapy: not allowed | | |
| | • Use of other therapy: not allowed | | |
| Kolb 1998 [79] | | • Proportion of patients with disease progression | |
| Study Design | Participants | Outcomes | Notes |
| • Type of Study: retrospective cohort | • Number: 18 | • Proportion of mortality | |
| • Trial Design: single centre | • Definition of diagnosis: clinical and radiological criteria: breathlessness, presence of fine crackles, especially in the bases, diffuse interstitial shadowing on chest radiography | • Idiopathic pulmonary fibrosis-related mortality | |
| • Country/ies: Germany | | • Proportion of patients with disease progression | |
| Study Duration | Participants | Outcomes | Notes |
| At least 1 year | • | | |
| | | • | |
| 163 |
• Age: median 58.5 years (range 36-75)
• Gender: 44% male
• Ethnicity: not stated
• Smoking status: 28% ever smoker
• Use of home oxygen therapy: not stated
• Time since diagnosis: median 19.5 months (range 6-90)
• Percentage of patients with surgical lung biopsy: not stated
• Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
• Lung function results (% predicted): mean FVC 52.39 (standard deviation 14.48)
• Resting oxyhaemoglobin saturation: not stated
• 6-minute walk distance: not stated
• Symptom assessment: not stated
• Use of systemic corticosteroid therapy: 100%
• Use of other therapy: cyclophosphamide 100%

Outcomes
Proportion of mortality
Duration of survival

Notes
Funding source: unclear

Kondoh 2005 [80]

Study Design
• Type of Study: retrospective cohort
• Trial Design: national multicentre (n = 6)
• Country/ies: Japan

Study Duration
4 years

Participants
• Number: 27
• Definition of diagnosis: the 2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment International Consensus Statement; the 2002 American Thoracic Society/European Respiratory Society International Multidisciplinary Consensus Classification of the Idiopathic Interstitial Pneumonias Statement
• Age: mean 56 years (standard deviation 10.9)
• Gender: 74% male
• Smoking status: 74% ever smoker
• Use of home oxygen therapy: not stated
• Time since diagnosis: not stated
• Percentage of patients with surgical lung biopsy: 100%
• Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
• Lung function results (% predicted): mean FVC 66.2 (standard deviation 18.2); mean DLCO 60.8 (standard deviation 18.2)
• Resting oxyhaemoglobin saturation: not stated
• 6-minute walk distance: not stated
• Symptom assessment: not stated
• Use of systemic corticosteroid therapy: 100%
• Use of other therapy: not stated

Outcomes
• Proportion of mortality
• Proportion of patients with disease progression

Notes
Funding source: unclear

Kondoh 2005

Study Design
Retrospective cohort study

Participants
27 patients

Definition of diagnosis
2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment International Consensus Statement; 2002 American Thoracic Society/European Respiratory Society International Multidisciplinary Consensus Classification of the Idiopathic Interstitial Pneumonias Statement

Age
Mean 56 years (SD 10.9)

Gender
74% male

Smoking status
74% ever smoker

Use of home oxygen therapy
Not stated

Time since diagnosis
Not stated

Percentage of patients with surgical lung biopsy
100%

Percentage of patients with definite usual interstitial pneumonia patterns on HRCT
Not stated

Lung function results (% predicted)
Mean FVC 66.2 (SD 18.2); mean DLCO 60.8 (SD 18.2)

Resting oxyhaemoglobin saturation
Not stated

6-minute walk distance
Not stated

Symptom assessment
Not stated

Use of systemic corticosteroid therapy
100%

Use of other therapy
Not stated

Outcomes
Proportion of mortality
Duration of survival

Notes
Funding source: unclear
### Kondoh 2010 [81]

**Study Design**
- Type of Study: retrospective cohort
- Trial Design: single centre
- Country/ies: Japan

**Study Duration**
- 3 years

**Participants**
- Number: 74
- Definition of diagnosis: the 2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment International Consensus Statement
- Age: mean 64.1 years (standard deviation 7.4)
- Gender: 82% male
- Ethnicity: not stated
- Smoking status: 73% ever smokers
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: 41%
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results (% predicted): mean FVC 77.0 (standard deviation 19.2); mean DLCO 59.3 (standard deviation 18.7)
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: not stated
- Use of other therapy: not stated

**Outcomes**
- Duration of survival

**Notes**
- Funding source: public

### Korthagen 2011 [82]

**Study Design**
- Type of Study: prospective cohort
- Trial Design: single centre
- Country/ies: Netherlands

**Study Duration**
- At least 1 year

**Participants**
- Number: 85
- Definition of diagnosis: the 2002 American Thoracic Society/European Respiratory Society International Multidisciplinary Consensus Classification of the Idiopathic Interstitial Pneumonias Statement
- Age: mean 65 years (standard deviation 10)
- Gender: 84% male
- Ethnicity: not stated
- Smoking status: 68% ever smoker
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: not stated
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results (% predicted): mean FEV1 82 (standard deviation 23); FVC 78 (standard deviation 23); mean DLCO 48 (standard deviation 18)
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: not stated
| Study Design | Study Duration | Participants | Outcomes | Notes |
|--------------|----------------|--------------|----------|-------|
| **Kotecha 2016 [83]** | At least 1 year | Number: 27 | Proportion of mortality, Progression-free survival | Funding source: unclear |
| **Study Design** | | Type of Study: prospective cohort | | |
| | | Trial Design: single centre | | |
| | | Country/ies: United Kingdom | | |
| **Participants** | | Number: 27 | | |
| | | Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management | | |
| | | Age: mean 72.8 years (standard deviation 9.5) | | |
| | | Gender: 85% male | | |
| | | Ethnicity: not stated | | |
| | | Smoking status: 70% ex-smokers | | |
| | | Use of home oxygen therapy: 7% | | |
| | | Time since diagnosis: mean 35 months (standard deviation 27 months) | | |
| | | Percentage of patients with surgical lung biopsy: not stated | | |
| | | Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated | | |
| | | Lung function results (% predicted): mean FVC 71.8 (standard deviation 18.1); mean DLCO 43.3 (standard deviation 16.0) | | |
| | | Resting oxyhaemoglobin saturation: not stated | | |
| | | 6-minute walk distance: not stated | | |
| | | Symptom assessment: not stated | | |
| | | Use of systemic corticosteroid therapy: not stated | | |
| | | Use of other therapy: not stated | | |
| **Outcomes** | | Proportion of mortality | | |
| | | Progression-free survival | | |
| **Notes** | | Funding source: unclear | | |

| Study Design | Study Duration | Participants | Outcomes | Notes |
|--------------|----------------|--------------|----------|-------|
| **Kreuter 2016 [84]** | At least 1 year | Number: 272 | | |
| **Study Design** | | Type of Study: retrospective cohort | | |
| | | Trial Design: single centre | | |
| | | Country/ies: Germany | | |
| **Study Duration** | | At least 1 year | | |
| **Participants** | | Number: 272 | | |
| | | Definition of diagnosis: multidisciplinary diagnosis | | |
| | | Age: mean 68.5 years (standard deviation 9) | | |
| | | Gender: 76.5% male | | |
| | | Ethnicity: not stated | | |
| | | Smoking status: 62% | | |
| | | Use of home oxygen therapy: not stated | | |
| | | Time since diagnosis: not stated | | |
| | | Percentage of patients with surgical lung biopsy: not stated | | |
| | | Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated | | |
| | | Lung function results (% predicted): mean FEV1 80.3 (standard deviation 18.0) | | |

| Notes | | Funding source: unclear | | |
| Study Design |  
| --- | --- |
| Kurashima 2010 [85] |  
| Type of Study: retrospective cohort  
| Trial Design: single centre  
| Country/ies: Japan |  
| Study Duration | At least 1 year |
| Participants |  
| Number: 660  
| Definition of diagnosis: Usual interstitial pneumonia pattern on HRCT and exclusion of all other diagnoses  
| Age: mean 72.9 years (standard deviation 8.1) for UIP group, 71.1 (7.7) for UIP/emphysema group  
| Gender: 83% male  
| Ethnicity: not stated  
| Smoking status: 83% ever smoker  
| Use of home oxygen therapy: not stated  
| Time since diagnosis: not stated  
| Percentage of patients with surgical lung biopsy: not stated  
| Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated  
| Lung function results (% predicted): mean FEV\(_1\) 86.7 (standard deviation 21.3) for UIP group, 87.9 (19.9) for UIP/emphysema group; mean FVC 71.8 (standard deviation 19.4) for UIP group, 87.1 (17.0) for UIP/emphysema group; mean DLCO 74.3 (standard deviation 20.1) for UIP group, 65.2 (20.9) for UIP/emphysema group  
| Resting oxyhaemoglobin saturation: not stated  
| 6-minute walk distance: not stated  
| Symptom assessment: not stated  
| Use of systemic corticosteroid therapy: 1.5%  
| Use of other therapy: not stated |  
| Outcomes | Duration of survival |
| Notes | Funding source: unclear |

| Study Design |  
| --- | --- |
| Le Rouzic 2015 [90] |  
| Type of Study: Retrospective cohort  
| Trial Design: Single centre  
| Country/ies: France |  
| Study Duration | At least 4 years |
| Participants |  
| Number: 66 (26 definite usual interstitial pneumonia)  
| Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis |
### Participants
- **Number:** 2635
- **Definition of diagnosis:** determined by transplant physicians
- **Age:** 36-54 years
- **Gender:** 62% male
- **Ethnicity:** 86% Caucasians 86%, African Americans 11%, Hispanic 6.56%
- **Smoking status:** not stated
- **Use of home oxygen therapy:** not stated
- **Time since diagnosis:** not stated
- **Percentage of patients with surgical lung biopsy:** not stated
- **Percentage of patients with definite usual interstitial pneumonia patterns on HRCT:** not stated
- **Lung function results (% predicted):** mean FEV<sub>1</sub> 53 (standard deviation 17) in Caucasians, 45 (17) in African Americans, 47 (17) in Hispanics; mean FVC 51 (standard deviation 17) in Caucasians, 44 (15) in African Americans, 45 (16) in Hispanics
- **Resting oxyhaemoglobin saturation:** not stated
- **6-minute walk distance:** not stated
- **Symptom assessment:** not stated

### Outcomes
- **Duration of survival**
- **Change in 6-minute walk distance**

### Notes
- **Funding source:** unclear

### Study Design
- **Type of Study:** retrospective cohort
- **Trial Design:** national multicentre (n = 94)
- **Country/ies:** USA

### Study Duration
- **At least 1 year**
| Study Design | Participants |
|--------------|--------------|
| Type of Study: retrospective cohort | Number: 86 |
| Trial Design: single centre | Definition of diagnosis: the 2002 American Thoracic Society/European Respiratory Society International Multidisciplinary Consensus Classification of the Idiopathic Interstitial Pneumonias Statement |
| Country/ies: Korea | Age: mean 61.3 years (standard deviation 8.9) |
| | Gender: 64% male |
| | Ethnicity: not stated |
| | Smoking status: not stated |
| | Use of home oxygen therapy: not stated |
| | Time since diagnosis: not stated |
| | Percentage of patients with surgical lung biopsy: not stated |
| | Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated |
| | Lung function results: not stated |
| | Resting oxyhaemoglobin saturation: not stated |
| | 6-minute walk distance: not stated |
| | Symptom assessment: not stated |
| | Use of systemic corticosteroid therapy: 83% |
| | Use of other therapy: not stated |

| Outcomes | Proportion of mortality |
| Notes | Funding source: unclear |
| Author provided additional data for clarification of study duration and survival/mortality. |

Lee 2012 [88]

| Study Design | Participants |
|--------------|--------------|
| Type of Study: Retrospective cohort | Number: 101 |
| Trial Design: Single centre | Definition of diagnosis: biopsy confirmed |
| Country/ies: Korea | Age: mean 59 years (standard deviation 7) |
| | Gender: 51% male |
| | Ethnicity: not stated |
| | Smoking status: not stated |
| | Use of home oxygen therapy: not stated |
| | Time since diagnosis: not stated |
| | Percentage of patients with surgical lung biopsy: not stated |
| | Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated |
| | Lung function results (% predicted): mean FEVC 82 (standard deviation |
| Study Design | • Type of Study: Retrospective cohort  
• Trial Design: Single centre  
• Country/ies: Korea |
|-------------|---------------------------------------------------------------|
| Study Duration | 3 years |
| Participants | • Number: 606  
• Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement:  
  Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management  
• Age: mean 69.5 years (standard deviation 8.6) for usual interstitial pneumonia group, 69.5 (10.3) for possible usual interstitial pneumonia group  
• Gender: 75% male for usual interstitial pneumonia group, 64.5% for possible usual interstitial pneumonia group  
• Ethnicity: not stated  
• Smoking status: 54% ever smokers for usual interstitial pneumonia group, 58% for possible usual interstitial pneumonia group  
• Use of home oxygen therapy: not stated  
• Time since diagnosis: not stated  
• Percentage of patients with surgical lung biopsy: not stated  
• Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated  
• Lung function results (% predicted): mean FEV1 90.8 (standard deviation 23.0) for usual interstitial pneumonia group, 94.9 (23.2) for possible usual interstitial pneumonia group; mean FVC 80.8 (standard deviation 20.4) for usual interstitial pneumonia group, 86.3 (17.6) for possible usual interstitial pneumonia group; mean DLCO 72.1 (standard deviation 23.7) for usual interstitial pneumonia group, 85.8 (26.5) for possible usual interstitial pneumonia group  
• Resting oxyhaemoglobin saturation: mean 98.0 % (standard deviation 4.0) for usual interstitial pneumonia group, 94.0% (2.5) for possible usual interstitial pneumonia group  
• 6-minute walk distance: not stated  
• Symptom assessment: mean MMRC scale 1.3 (standard deviation 1.2) for usual interstitial pneumonia group, 1.0 (1.0) for possible usual interstitial pneumonia group  
• Use of systemic corticosteroid therapy: not stated  
• Use of other therapy: not stated |
| Outcomes | Proportion of mortality |
| Notes | Funding source: unclear |

**Lee 2015 [89]**

| Study Design | • Type of Study: Retrospective cohort |
|-------------|-------------------------------------|
| Study Duration | 3 years |
| Participants | • Number: 606  
• Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement:  
  Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management  
• Age: mean 69.5 years (standard deviation 8.6) for usual interstitial pneumonia group, 69.5 (10.3) for possible usual interstitial pneumonia group  
• Gender: 75% male for usual interstitial pneumonia group, 64.5% for possible usual interstitial pneumonia group  
• Ethnicity: not stated  
• Smoking status: 54% ever smokers for usual interstitial pneumonia group, 58% for possible usual interstitial pneumonia group  
• Use of home oxygen therapy: not stated  
• Time since diagnosis: not stated  
• Percentage of patients with surgical lung biopsy: not stated  
• Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated  
• Lung function results (% predicted): mean FEV1 90.8 (standard deviation 23.0) for usual interstitial pneumonia group, 94.9 (23.2) for possible usual interstitial pneumonia group; mean FVC 80.8 (standard deviation 20.4) for usual interstitial pneumonia group, 86.3 (17.6) for possible usual interstitial pneumonia group; mean DLCO 72.1 (standard deviation 23.7) for usual interstitial pneumonia group, 85.8 (26.5) for possible usual interstitial pneumonia group  
• Resting oxyhaemoglobin saturation: mean 98.0 % (standard deviation 4.0) for usual interstitial pneumonia group, 94.0% (2.5) for possible usual interstitial pneumonia group  
• 6-minute walk distance: not stated  
• Symptom assessment: mean MMRC scale 1.3 (standard deviation 1.2) for usual interstitial pneumonia group, 1.0 (1.0) for possible usual interstitial pneumonia group  
• Use of systemic corticosteroid therapy: not stated  
• Use of other therapy: not stated |
| Outcomes | Proportion of mortality |
| Notes | Funding source: unclear |

**Li 2010 [91]**

| Study Design | • Type of Study: Retrospective cohort |
|-------------|-------------------------------------|
### Study Design
- **Type of Study**: prospective cohort
- **Trial Design**: single centre
- **Country/ies**: China

### Study Duration
- **At least 1 year**

### Participants
- **Number**: 126
- **Definition of diagnosis**: the 2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment International Consensus Statement
- **Age**: not stated
- **Gender**: not stated
- **Ethnicity**: not stated
- **Smoking status**: 60% ever smokers
- **Use of home oxygen therapy**: not stated
- **Time since diagnosis**: not stated
- **Percentage of patients with surgical lung biopsy**: not stated
- **Percentage of patients with definite usual interstitial pneumonia patterns on HRCT**: 30%
- **Lung function results**: mean DLCO 55% predicted
- **Resting oxyhaemoglobin saturation**: not stated
- **6-minute walk distance**: not stated
- **Symptom assessment**: not stated
- **Use of systemic corticosteroid therapy**: 75%
- **Use of other therapy**: not stated

### Outcomes
- **Duration of survival**

### Notes
- **Funding source**: unclear

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### Li 2012 [92]

| Study Design | Type of Study: prospective cohort |
|--------------|----------------------------------|
|              | Trial Design: single centre      |
|              | Country/ies: China               |
| Study Duration | 1 year                           |

| Participants | Number: 30 |
|--------------|------------|
| Definition of diagnosis: ATS/ERS guideline (specific guideline not stated) |
| Age: mean 65 +/- 10 years       |
| Gender: 53% male |
| Ethnicity: not stated |
| Smoking status: 33% ever smoker |
| Use of home oxygen therapy: not stated |
| Time since diagnosis: not stated |
| Percentage of patients with surgical lung biopsy: not stated |
| Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated |
| Lung function results: not stated |
| Resting oxyhaemoglobin saturation: not stated |
| 6-minute walk distance: not stated |
| Symptom assessment: St George’s Respiratory Questionnaire score: mean 61.7 +/- 17.72 |
| Use of systemic corticosteroid therapy: not stated |
| Use of other therapy: not stated |

| Outcomes | Proportion of mortality |
|----------|-------------------------|
|          | Duration of survival    |

| Notes | Funding source: unclear |
|-------|-------------------------|
### Li 2015 [93]

| **Study Design** | • Type of Study: retrospective cohort  
| | • Trial Design: single centre  
| | • Country/ies: China  
| **Study Duration** | mean 46 months (range 13 to 84)  
| **Participants** | • Number: 55  
| | • Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management  
| | • Age: mean 60 years  
| | • Gender: 71% male  
| | • Ethnicity: not stated  
| | • Smoking status: 42% ever smoker  
| | • Use of home oxygen therapy: not stated  
| | • Time since diagnosis: not stated  
| | • Percentage of patients with surgical lung biopsy: not stated  
| | • Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated  
| | • Lung function results: not stated  
| | • Resting oxyhaemoglobin saturation: not stated  
| | • 6-minute walk distance: not stated  
| | • Symptom assessment: not stated  
| | • Use of systemic corticosteroid therapy: not stated  
| | • Use of other therapy: not stated  
| **Outcomes** | Duration of survival  
| **Notes** | Funding source: unclear  

### Lindell 2015 [94]

| **Study Design** | • Type of Study: retrospective cohort  
| | • Trial Design: single centre  
| | • Country/ies: United States  
| **Study Duration** | At least 1 year  
| **Participants** | • Number: 404  
| | • Definition of diagnosis: diagnosis at interstitial lung disease centres  
| | • Age: mean 71.5 years (standard deviation 9.2 years)  
| | • Gender: 65% male  
| | • Ethnicity: Caucasian 97.3%  
| | • Smoking status: not stated  
| | • Use of home oxygen therapy: not stated  
| | • Time since diagnosis: not stated  
| | • Percentage of patients with surgical lung biopsy: not stated  
| | • Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated  
| | • Lung function results (% predicted): mean FVC 60.5 (standard deviation 18.1); mean DLCO 41.2 (standard deviation 16.7)  
| | • Resting oxyhaemoglobin saturation: not stated  
| | • 6-minute walk distance: not stated  
| | • Symptom assessment: not stated  
| | • Use of systemic corticosteroid therapy: not stated  
| | • Use of other therapy: not stated  
| **Outcomes** | • Proportion of mortality  

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### Liu 2017 [95]

| Study Design | Type of Study: retrospective cohort  
|             | Trial Design: single centre  
|             | Country/ies: China  
| Study Duration | 1 year  
| Participants | Number: 69  
|             | Definition of diagnosis: the 2002 American Thoracic Society/European Respiratory Society International Multidisciplinary Consensus Classification of the Idiopathic Interstitial Pneumonias Statement  
|             | Age: mean 68 years (standard deviation 6)  
|             | Gender: 67% male  
|             | Ethnicity: not stated  
|             | Smoking status: 72% ever smokers  
|             | Use of home oxygen therapy: not stated  
|             | Time since diagnosis: not stated  
|             | Percentage of patients with surgical lung biopsy: not stated  
|             | Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated  
|             | Lung function results (% predicted): mean FVC 71 (standard deviation 15), mean DLCO 52 (standard deviation 15)  
|             | Resting oxyhaemoglobin saturation: not stated  
|             | 6-minute walk distance: not stated  
|             | Symptom assessment: not stated  
|             | Use of systemic corticosteroid therapy: not stated  
|             | Use of other therapy: not stated  
| Outcomes | Duration of survival (Kaplan-Meier Curve – data not extractable)  
| Notes | Duration of survival  

### Lutherer 2011 [96]

| Study Design | Type of Study: prospective cohort  
|             | Trial Design: single centre  
|             | Country/ies: United States  
| Study Duration | 1 year  
| Participants | Number: 12  
|             | Definition of diagnosis: ATS criteria (no further details specified)  
|             | Age: mean 67 years (range 50-82)  
|             | Gender: not stated  
|             | Ethnicity: not stated  
|             | Smoking status: not stated  
|             | Use of home oxygen therapy: not stated  
|             | Time since diagnosis: not stated  
|             | Percentage of patients with surgical lung biopsy: 25%  
|             | Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated  
|             | Lung function results (% predicted): mean FVC 57 (range 36.7-73.4)  
|             | Resting oxyhaemoglobin saturation: not stated  
|             | 6-minute walk distance: not stated  
|             | Symptom assessment: not stated  
| Notes | Funding source: unclear
Malouf 2011 [181]

**Study Design**
- Type of Study: randomised controlled trial
- Trial Design: parallel-group, national multicentre (n = 6)
- Country/ies: Australia

**Study Duration**
- 3 years

**Participants**
- Number: 45 (placebo group)
- Definition of diagnosis: the 2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment International Consensus Statement
- Age: mean 60 years (standard deviation 9)
- Gender: 71% male
- Ethnicity: not stated
- Smoking status: not stated
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: 100%
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results (% predicted): mean FVC 69 (standard deviation 20); mean DLCO%: 42 (standard deviation 14)
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: placebo: mean 451m (standard deviation 118)
- Symptom assessment: Medical Outcomes Study Short-Form Health Survey (SF-36) – physical component: mean 48.60 (standard deviation 12.64); Medical Outcomes Study Short-Form Health Survey (SF-36) – mental component: mean 28.35 (standard deviation 9.91)
- Use of systemic corticosteroid therapy: 26.7%
- Use of other therapy: not stated

**Outcomes**
- Proportion of mortality

**Notes**
- Funding source: unclear

Mapel 1998 [97]

**Study Design**
- Type of Study: prospective cohort
- Trial Design: national using the New Mexico Interstitial Lung Disease Registry
- Country/ies: USA

**Study Duration**
- At least 1 year

**Participants**
- Number: 209
- Definition of diagnosis: International Classification of Diseases, 9th revision
- Age: mean 71.7 years (standard deviation 12.3)
- Gender: 55% male
- Ethnicity: Non-Hispanic White 61.2%, Hispanic 23.0%, Native American 1.9%, African American 1.4%, Other 12.4%
- Smoking status: 61% ever smoker, 10.5% unknown
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
### Martinez 2014 [182]

| Study Design | Study Duration | Participants | Outcomes |
|--------------|----------------|--------------|----------|
| Type of Study: randomised controlled trial | 60 weeks | Number: 131 (placebo group) | Proportion of mortality |
| Trial Design: phase 3, parallel-group, multicentre (n = 25) | | Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management | Respiratory-specific mortality |
| Country/ies: United States | | Age: mean 67.2 years (standard deviation 8.2) | Change in forced vital capacity |
| | | Gender: 74.8% male | Change in diffusing capacity for carbon monoxide |
| | | Ethnicity: Caucasian 96.2% | Change in dyspnoea (University of California San Diego Shortness of Breath Questionnaire) |
| | | Smoking status: 74.8% ever smoker | Change in health-related quality of life (St George’s Respiratory Questionnaire, EuroQoL visual analogue score) |
| | | Use of home oxygen therapy: not stated | Change in 6-minute walk distance |
| | | Time since diagnosis: mean 1.1 years (standard deviation 1.0) | |
| | | Percentage of patients with surgical lung biopsy: not stated | |
| | | Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated | |
| | | Lung function results (% predicted): mean FVC 73.4 (standard deviation 14.3); mean DLCO 46.0 (standard deviation 12.2) | |
| | | Resting oxyhaemoglobin saturation: not stated | |
| | | 6-minute walk distance: mean 375m (standard deviation 105) | |
| | | Symptom assessment: University of California San Diego Shortness of Breath Questionnaire: mean 27.1 (standard deviation 18.7); total St George’s Respiratory Questionnaire: mean 38.0 (standard deviation 17.2); Short Form- physical score: mean 40.7 (standard deviation 9.3); Short Form-36 mental score: 55.3 (standard deviation 7.5); EuroQoL Visual Analogue Scale: mean 77.7 (standard deviation 14.3) | |
| | | Use of systemic corticosteroid therapy: 0% | |
| | | Use of other therapy: not stated | |

Notes:
- Funding source: public

Outcomes:
- Proportion of mortality
- Respiratory-specific mortality
- Change in forced vital capacity
- Change in diffusing capacity for carbon monoxide
- Change in dyspnoea (University of California San Diego Shortness of Breath Questionnaire)
- Change in health-related quality of life (St George’s Respiratory Questionnaire, EuroQoL visual analogue score)
- Change in 6-minute walk distance
### Marulli 2010 [98]

| **Study Design** | • Type of Study: Retrospective cohort  
|                  | • Trial Design: single centre  
|                  | • Country/ies: Italy  
| **Study Duration** | 1 year  
| **Participants** | • Number: 56  
|                  | • Definition of diagnosis: unclear  
|                  | • Age: not stated  
|                  | • Gender: not stated  
|                  | • Ethnicity: not stated  
|                  | • Smoking status: not stated  
|                  | • Use of home oxygen therapy: not stated  
|                  | • Time since diagnosis: not stated  
|                  | • Percentage of patients with surgical lung biopsy: not stated  
|                  | • Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated  
|                  | • Lung function results: median DLCO 14.9 +/- 7.4 for non-survivors, 25.3 +/- 12.3 for survivors  
|                  | • Resting oxyhaemoglobin saturation: not stated  
|                  | • 6-minute walk distance: not stated  
|                  | • Symptom assessment: not stated  
|                  | • Use of systemic corticosteroid therapy: not stated  
|                  | • Use of other therapy: not stated  
| **Outcomes** | Proportion of mortality  
| **Notes** | Funding source: mixed

### Mason 2007 [99]

| **Study Design** | • Type of Study: retrospective cohort  
|                  | • Trial Design: single centre  
|                  | • Country/ies: United States  
| **Study Duration** | 1 year  
| **Participants** | • Number: 82  
|                  | • Definition of diagnosis: the 2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment International Consensus Statement  
|                  | • Age: mean 52 years (standard deviation 11)  
|                  | • Gender: 63% male  
|                  | • Ethnicity: Caucasian 84%  
|                  | • Smoking status: 59% ever smoker  
|                  | • Use of home oxygen therapy: not stated  
|                  | • Time since diagnosis: not stated  
|                  | • Percentage of patients with surgical lung biopsy: not stated  
|                  | • Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated  
|                  | • Lung function results (% predicted): mean FEV1 44 (standard deviation 15); mean FVC 44 (standard deviation 15)  
|                  | • Resting oxyhaemoglobin saturation: not stated  
|                  | • 6-minute walk distance: not stated  
|                  | • Symptom assessment: not stated  
|                  | • Use of systemic corticosteroid therapy: not stated  
| **Notes** | Funding source: unclear
McKeown 2009 [100]

**Study Design**
- Type of Study: prospective cohort
- Trial Design: single centre trial
- Country/ies: United Kingdom

**Study Duration**
- 3 years

**Participants**
- Number: 20
- Definition of diagnosis: the 2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment International Consensus Statement
- Age: mean 70 years (range 52-86)
- Gender: 80% male
- Ethnicity: not stated
- Smoking status: 85% ever smoker
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: 25%
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results (% predicted): mean FVC 73 (standard deviation 19.7); mean DLCO 49 (standard deviation 14.4)
- Resting oxyhaemoglobin saturation: mean 94% (standard deviation 3)
- 6-minute walk distance: not stated
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: not stated
- Use of other therapy: not stated

**Outcomes**
- Proportion of mortality
- Duration of survival

**Notes**
- Funding source: unclear

Meier-Sydow 1979 [101]

**Study Design**
- Type of Study: retrospective cohort
- Trial Design: single centre
- Country/ies: Germany

**Study Duration**
- At least 1 year

**Participants**
- Number: 21
- Definition of diagnosis: histologic pattern of UIP or based on history, clinical findings on physical examination, physiologic data and radiographic findings
- Age: not stated
- Gender: not stated
- Ethnicity: not stated
- Smoking status: not stated
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: not stated
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results: mean FVC 1.73 (standard deviation 0.52) for
|                      | azathioprine group, 2.32 (0.88) for D-penicillamine group |
|----------------------|----------------------------------------------------------|
|                      | • Resting oxyhaemoglobin saturation: not stated          |
|                      | • 6-minute walk distance: not stated                     |
|                      | • Symptom assessment: not stated                         |
|                      | • Use of systemic corticosteroid therapy: 100%           |
|                      | • Use of other therapy: azathioprine 52%, D-penicillamine 48% |

**Outcomes**
Proportion of mortality

**Notes**
Funding source: unclear

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**Meier-Sydow 1990 [102]**

**Study Design**
- Type of Study: prospective cohort
- Trial Design: single centre
- Country/ies: Germany

**Study Duration**
10 years

**Participants**
- Number: 37
- Definition of diagnosis: lung biopsy
- Age: mean 52 +/- 11 years for prednisolone group, 58 +/- 9 for azathioprine group, 53 +/- 11 for D-penicillamine group
- Gender: not stated
- Ethnicity: not stated
- Smoking status: not stated
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: not stated
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results: mean FVC 2.7 +/- 1.0 L for prednisolone group, 2.2 +/- 0.7 for azathioprine group, 2.0 +/- 0.5 for D-penicillamine group; mean DLCO 9.6 +/- 5.2 for prednisolone group, 9.3 +/- 2.8 for azathioprine group, 7.4 +/- 3.9 for D-penicillamine group
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: 30%
- Use of other therapy: azathioprine 41%, D-penicillamine 30%

**Outcomes**
Duration of survival

**Notes**
Funding source: unclear

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**Mermigkis 2015 [103]**

**Study Design**
- Type of Study: prospective cohort
- Trial Design: single centre
- Country/ies: Greece

**Study Duration**
2 years

**Participants**
- Number: 55
- Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management
- Age: not stated
- Gender: not stated
- Ethnicity: not stated
- Smoking status: not stated
- Use of home oxygen therapy: not stated
| Outcomes | Proportion of mortality |
|----------|-------------------------|
| Notes    | Funding source: unclear |

**Mirrani 2012 [104]**

| Study Design |
|--------------|
| Type of Study: retrospective cohort |
| Trial Design: single centre |
| Country/ies: United Kingdom |

| Study Duration |
|----------------|
| 10 years |

| Participants |
|---------------|
| Number: 72 |
| Definition of diagnosis: unclear |
| Age: not stated |
| Gender: not stated |
| Ethnicity: not stated |
| Smoking status: not stated |
| Use of home oxygen therapy: not stated |
| Time since diagnosis: not stated |
| Percentage of patients with surgical lung biopsy: 2.7% |
| Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated |
| Lung function results (% predicted): mean FVC 84 (standard deviation 20.9); mean DLCO 52 (standard deviation 20) |
| Resting oxyhaemoglobin saturation: not stated |
| 6-minute walk distance: not stated |
| Symptom assessment: not stated |
| Use of systemic corticosteroid therapy: not stated |
| Use of other therapy: not stated |

| Outcomes |
|----------|
| Proportion of mortality |
| Duration of survival |

| Notes |
|-------|
| Funding source: unclear |

**Moeller 2009 [105]**

| Study Design |
|--------------|
| Type of Study: prospective cohort |
| Trial Design: single centre |
| Country/ies: Canada |

| Study Duration |
|----------------|
| 2 years |

| Participants |
|---------------|
| Number: 58 |
| Definition of diagnosis: the 2002 American Thoracic Society/European |
Respiratory Society International Multidisciplinary Consensus Classification of the Idiopathic Interstitial Pneumonias Statement

- Age: mean 68 years (standard error 9.8) for stable patients, 72.4 years (6) for acute patients
- Gender: 33% male
- Ethnicity: not stated
- Smoking status: not stated
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: 29%
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results (% predicted): mean FVC 65.3 (standard error 18.1) for stable patients, 56.3 (17.6) for acute patients
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: not stated
- Use of other therapy: not stated

Outcomes
- Proportion of mortality
- Duration of survival

Notes
- Funding source: public

Mogulkoc 2001 [106]

| Study Design | Type of Study: prospective cohort |
|--------------|----------------------------------|
|              | Trial Design: single centre       |
|              | Country/ies: UK                  |

| Study Duration | 2 years |

| Participants   | Number: 95 |
|----------------|------------|
|                | Definition of diagnosis: exclusion of other diagnosis of interstitial lung disease including collagen vascular disease, allergic extrinsic alveolitis |
|                | Age: mean 55.5 years (standard deviation 8.4) |
|                | Gender: 71% male |
|                | Ethnicity: not stated |
|                | Smoking status: 61% ever smoker |
|                | Use of home oxygen therapy: not stated |
|                | Time since diagnosis: not stated |
|                | Percentage of patients with surgical lung biopsy: 44% |
|                | Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated |
|                | Lung function results (% predicted): mean FEV1 70 (standard deviation 22); mean FVC 72 (standard deviation 25); mean DLCO 49 (standard deviation 17) |
|                | Resting oxyhaemoglobin saturation: not stated |
|                | 6-minute walk distance: not stated |
|                | Symptom assessment: not stated |
|                | Use of systemic corticosteroid therapy: not stated |
|                | Use of other therapy: not stated |

| Outcomes       | Proportion of mortality |
|----------------|-------------------------|
| Notes          | Funding source: unclear |

Moore 2013 [107]
### Study Design
- Type of Study: retrospective cohort
- Trial Design: single centre
- Country/ies: United Kingdom

### Study Duration
1 year

### Participants
- Number: 58
- Definition of diagnosis: unclear
- Age: mean 80.2 years for fast progressors, 71.8 for slow progressors
- Gender: 69% male
- Ethnicity: not stated
- Smoking status: 40% ever smokers for fast progressors, 60% for slow progressors
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: not stated
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results: not stated
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: not stated
- Use of other therapy: not stated

### Outcomes
- Duration of survival

### Notes
- Funding source: unclear

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### Morawiec 2011 [108]

| Study Design | Type of Study: prospective cohort |
|--------------|-----------------------------------|
|              | Trial Design: single centre       |
|              | Country/ies: France               |

### Study Duration
1 year

### Participants
- Number: 18
- Definition of diagnosis: unclear
- Age: median 67 years (interquartile range 65-72)
- Gender: 88.9% male
- Ethnicity: 94.4% Caucasian
- Smoking status: 50% ever smoker
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: not stated
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results (% predicted): mean FVC 70 (interquartile range 62-78); mean DLCO 42 (interquartile range 39-47)
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: mean 145m (interquartile range 350-425)
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: not stated
- Use of other therapy: not stated

### Outcomes
- Proportion of mortality

### Notes
- Funding source: unclear

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### Moua 2016 [109]
| Study Design          | • Type of Study: retrospective cohort  
|                      | • Trial Design: single centre  
|                      | • Country/ies: United States

| Study Duration       | At least 1 year

| Participants         | • Number: 100  
|                      | • Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management  
|                      | • Age: mean 73.1 years (standard deviation 8.7)  
|                      | • Gender: not stated  
|                      | • Smoking status: not stated  
|                      | • Use of home oxygen therapy: not stated  
|                      | • Time since diagnosis: not stated  
|                      | • Percentage of patients with surgical lung biopsy: not stated  
|                      | • Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated  
|                      | • Lung function results: not stated  
|                      | • Resting oxyhaemoglobin saturation: not stated  
|                      | • 6-minute walk distance: not stated  
|                      | • Symptom assessment: not stated  
|                      | • Use of systemic corticosteroid therapy: not stated  
|                      | • Use of other therapy: not stated

| Outcomes             | Duration of survival

| Notes                | Funding source: public

Mura 2012 [110]

| Study Design          | • Type of Study: prospective and retrospective cohort  
|                      | • Trial Design: single centre  
|                      | • Country/ies: Italy

| Study Duration       | At least 1 year

| Participants         | • Number: 70 for prospective study, 68 for retrospective study  
|                      | • Definition of diagnosis: the 2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment International Consensus Statement  
|                      | • Age: mean 67 years (standard deviation 8) for prospective study, 62 (9) for retrospective study  
|                      | • Gender: 81% male for prospective study, 74% for retrospective study  
|                      | • Smoking status: 63% ever smokers for prospective study, 62% for retrospective study  
|                      | • Use of home oxygen therapy: not stated  
|                      | • Time since diagnosis: not stated  
|                      | • Percentage of patients with surgical lung biopsy: 33% for prospective study, 32% for retrospective study  
|                      | • Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated  
|                      | • Lung function results (% predicted): mean FVC 75 (standard deviation 22) for prospective study, 75 (21) for retrospective study; mean DLCO 46 (standard deviation 19) for prospective study, 55 (17) for retrospective study  
|                      | • Resting oxyhaemoglobin saturation: not stated
| Nadrous 2004 [111] |
|---------------------|
| **Study Design**    |
| Type of Study: retrospective cohort |
| Trial Design: single centre |
| Country/ies: United States |
| **Study Duration** 2 years |
| **Participants**    |
| Number: 478 |
| Definition of diagnosis: compatible clinical characteristics plus either consistent high-resolution chest CT findings or histopathologic evidence |
| Age: not stated |
| Gender: 70% male |
| Ethnicity: not stated |
| Smoking status: 66% ever smokers, 1% unknown |
| Use of home oxygen therapy: 16% |
| Time since diagnosis: not stated |
| Percentage of patients with surgical lung biopsy: not stated |
| Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated |
| Lung function results (% predicted): mean FEV1 70.6 (standard deviation 17.4); mean FVC 67.7 (standard deviation 18.8); mean DLCO 51.7 (standard deviation 16.4) |
| Resting oxyhaemoglobin saturation: mean 93.1% (standard deviation 2.4) |
| 6-minute walk distance: not stated |
| Symptom assessment: not stated |
| Use of systemic corticosteroid therapy: 26% |
| Use of other therapy: colchicine 38% |
| **Outcomes** Duration of survival |
| **Notes** Funding source: public |

| Nagai 1998 [112] |
|-------------------|
| **Study Design**  |
| Type of Study: retrospective cohort |
| Trial Design: national |
| Country/ies: Japan |
| **Study Duration** 7 years |
| **Participants** |
| Number: 64 |
| Definition of diagnosis: unclear |
| Age: mean 59.5 years (standard deviation 10) |
| Gender: 86% male |
| Ethnicity: not stated |
| Smoking status: 83% ever smoker |
| Use of home oxygen therapy: not stated |
| Time since diagnosis: not stated |
| Nambiar 2017 [113] |  |
|---|---|
| **Study Design** |  |
| • Type of Study: retrospective cohort  
• Trial Design: national multicentre (n = 3)  
• Country/ies: USA |  |
| **Study Duration** | 1 year |
| **Participants** |  |
| • Number: 41  
• Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management  
• Age: mean 66.1 years (standard deviation 8.9)  
• Gender: 63.4% male  
• Ethnicity: Caucasian 58%, Hispanic 39%  
• Smoking status: 78% ex-smoker  
• Use of home oxygen therapy: 85.4%  
• Time since diagnosis: mean 15.5 years (standard deviation 9.5)  
• Percentage of patients with surgical lung biopsy: 29.3%  
• Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: 90.2%  
• Lung function results (% predicted): mean FVC 63.3 (standard deviation 17.3); mean DLCO 44.1 (standard deviation 17.5)  
• Resting oxyhaemoglobin saturation: not stated  
• 6-minute walk distance: not stated  
• Symptom assessment: not stated  
• Use of systemic corticosteroid therapy: 39%  
• Use of other therapy: mycophenolate mofetil 27%, N-acetylcysteine 59%, azathioprine 15% |  |
| **Outcomes** |  |
| • Duration of survival  
• Proportion of patients with disease progression |  |
| **Notes** | Funding source: public |

| Natsuizaka 2014 [114] |  |
|---|---|
| **Study Design** |  |
| • Type of Study: retrospective cohort  
• Trial Design: national based on the application of the Certificate of Medical Benefit in Hokkaido prefecture  
• Country/ies: Japan |  |
| **Study Duration** | At least 1 year |
| **Participants** |  |
| • Number: 553  
• Definition of diagnosis: the 2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Management |  |
Treatment International Consensus Statement

- Age: mean 70 years (standard deviation 9)
- Gender: 73% male
- Ethnicity: not stated
- Smoking status: 68% ever smoker
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: not stated
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results: 61.1% FVC < 80% predicted, 84.3% DLCO < % predicted
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: not stated
- Use of other therapy: not stated

**Outcomes**
- Duration of survival

**Notes**
- Funding source: unclear

### Nicholson 2000 [115]

**Study Design**
- Type of Study: retrospective cohort
- Trial Design: single centre
- Country/ies: United Kingdom

**Study Duration**
- 10 years

**Participants**
- Number: 37
- Definition of diagnosis: bibasal/widespread crackles (most prominent at bases); abnormalities consistent with bilateral lung fibrosis on chest radiography; restrictive functional defect or isolated reduction DLCO; absence of occupational/environmental cause for pulmonary fibrosis
- Age: mean 57.2 years (standard deviation 7.1)
- Gender: 89% male
- Ethnicity: not stated
- Smoking status: 86% ever smoker
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: not stated
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results (% predicted): mean FVC 71.5 (standard deviation 16.1); mean DLCO 43.5 (standard deviation 11.6)
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: not stated
- Use of other therapy: not stated

**Outcomes**
- Proportion of mortality
- Duration of survival

**Notes**
- Funding source: unclear

### Nicol 2015 [116]

**Study Design**
- Type of Study: retrospective cohort
| Study Design | Type of Study: retrospective cohort
| Country/ies: Japan |
| Study Duration | 1 year |
| Participants | Number: 114
| Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management
| Age: mean 73.8 years (standard deviation 6.8)
| Gender: 82% male
| Ethnicity: not stated
| Smoking status: 75% ever smokers, 13% unknown
| Use of home oxygen therapy: not stated
| Time since diagnosis: not stated
| Percentage of patients with surgical lung biopsy: not stated
| Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
| Lung function results (% predicted): mean FEV1 77.2 (standard deviation 19.6); mean FVC 75.2 (standard deviation 23.3); mean DLCO 69.5 (standard deviation 22.7)
| Resting oxyhaemoglobin saturation: not stated
| 6-minute walk distance: not stated
| Outcomes | Proportion of mortality |
|---|---|
| Notes | • Funding source: no funding  
• Author provided additional data for clarification of study duration and survival/mortality. |

### Nishiyama 2016b [118]

| Study Design | • Type of Study: prospective cohort  
• Trial Design: single centre  
• Country/ies: Japan |
|---|---|
| Study Duration | At least 1 year |
| Participants | • Number: 44  
• Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management  
• Age: mean 72.3 years (standard deviation 7.2)  
• Gender: 80% male  
• Ethnicity: not stated  
• Smoking status: not stated  
• Use of home oxygen therapy: not stated  
• Time since diagnosis: not stated  
• Percentage of patients with surgical lung biopsy: not stated  
• Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated  
• Lung function results (% predicted): mean FEV1 84.4% (standard deviation 17.4); mean FVC 82.6 (standard deviation 20.8); mean DLCO 69.3 (standard deviation 19.3)  
• Resting oxyhaemoglobin saturation: not stated  
• 6-minute walk distance: mean 412m (standard deviation 90)  
• Symptom assessment: St George’s Respiratory Questionnaire-Symptoms score: mean 44.7 (standard deviation 22.8); St George’s Respiratory Questionnaire-Activity score: mean 45.9 (standard deviation 24.6); St George’s Respiratory Questionnaire-Impacts score: mean 26.6 (standard deviation 20.4); total St George’s Respiratory Questionnaire score: mean 35.7 (standard deviation 20.0); Hospital Anxiety and Depression Scale (Anxiety): mean 5.1 (standard deviation 3.8); Hospital Anxiety and Depression Scale (Depression): mean 6.0 (standard deviation 2.9)  
• Use of systemic corticosteroid therapy: not stated  
• Use of other therapy: not stated |
| Outcomes | Duration of survival |
| Notes | • Funding source: unclear  
• Author provided additional data for clarification of study duration and survival/mortality. |

### Noble 2011 [183]

| Study Design | • Type of Study: randomised controlled trial  
• Trial Design: phase 3, parallel-group, international multicentre (n = 110)  
• Country/ies: Australia, Belgium, Canada, France, Germany, Ireland, Italy, Mexico, Poland, Spain, Switzerland, United Kingdom, United States |
|---|---|
| Study Duration | 72 weeks |
**Participants**
- Number: 347
- Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management
- Age: mean 66.3 years (standard deviation 7.5) for Study 004, 67 (standard deviation 7.8) for Study 006
- Gender: 73% male
- Ethnicity: Caucasian 98%
- Smoking status: 67% ever smokers
- Use of home oxygen therapy: 21%
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: 52%
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: 93%
- Lung function results (% predicted): mean FVC 76.2 (standard deviation 15.5) for Study 004, 73.1 (14.2) for Study 006; mean DLCO 46.1 (standard deviation 10.2) for Study 004, 47.4 (9.2) for Study 006
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: mean 410 m (standard deviation 90.9) for Study 004, 399.1m (89.7) for Study 006
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: not stated
- Use of other therapy: not stated

**Outcomes**
- Proportion of mortality
- Idiopathic pulmonary fibrosis-related mortality
- Change in forced vital capacity
- Change in dyspnoea (University of California San Diego Shortness of Breath Questionnaire)
- Change in 6-minute walk distance
- Proportion of patients with disease progression

**Notes**
- Funding source: industry

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**Oda 2014 [119]**

**Study Design**
- Type of Study: retrospective cohort
- Trial Design: single centre
- Country/ies: Japan

**Study Duration**
- 1 year

**Participants**
- Number: 98
- Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management
- Age: mean 76 years (standard deviation 8.0)
- Gender: 56% male
- Ethnicity: not stated
- Smoking status: 70% ever smoker
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: not stated
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results (% predicted): mean FEV1 85.1 (standard deviation 9.15); mean FVC 71.0 (standard deviation 20.8); mean DLCO 60.0
### Oldham 2015 [120]

**Study Design**
- Type of Study: retrospective cohort
- Trial Design: single centre
- Country/ies: United States

**Study Duration**
- At least 1 year

**Participants**
- Number: 196
- Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management
- Age: mean 68.1 years (standard deviation 8.6)
- Gender: 74.5% male
- Ethnicity: Caucasian 80.1%, Hispanic 9.2%, African American 8.2%, Asian 2.5%
- Smoking status: 74% ever smoker
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: 79%
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results (% predicted): mean FVC 59.5 (standard deviation 12.2) for hypothyroidism group, 65.4 (18.1) for non-hypothyroidism group; mean DLCO 43.3 (standard deviation 16.3) for hypothyroidism group, 50.4 (17.6) for non-hypothyroidism group
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: 12.8%
- Use of other therapy: not stated

**Outcomes**
- Proportion of survival
- Proportion of patients with disease progression

**Notes**
- Funding source: public
- Author provided additional data for clarification of study duration and survival/mortality.

### Parker 2016 [184]

**Study Design**
- Type of Study: randomised controlled trial
- Trial Design: phase 2, parallel-group, international multicentre (n = 48)
- Country/ies: Australia, Canada, Israel, Peru, South Korea, United States

**Study Duration**
- 1 year

**Participants**
- Number: 57 (placebo group)
- Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management
• Age: mean 67.5 years (standard deviation 6.1)
• Gender: 78.9% male
• Ethnicity: White 75.4%, Asian 12.3%, other 12.3%
• Smoking status: not stated
• Use of home oxygen therapy: not stated
• Time since diagnosis: not stated
• Percentage of patients with surgical lung biopsy: not stated
• Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
• Lung function results (% predicted): mean FVC 70.3 (standard deviation 12.0); mean DLCO 47.0 (standard deviation 13.8)
• Resting oxyhaemoglobin saturation: not stated
• 6-minute walk distance: mean 391m (standard deviation 112)
• Symptom assessment: not stated
• Use of systemic corticosteroid therapy: not stated
• Use of other therapy: not stated

Outcomes
• Proportion of mortality
• Respiratory-specific mortality
• Change in forced vital capacity
• Change in diffusing capacity for carbon monoxide
• Change in dyspnoea (University of California San Diego Shortness of Breath Questionnaire)
• Change in health-related quality of life (St George’s Respiratory Questionnaire, EuroQol-5D)
• Change in 6-minute walk distance
• Proportion of patients with disease progression

Notes
Funding source: industry

Prasse 2009 [121]

Study Design
• Type of Study: prospective cohort
• Trial Design: international multicentre, number of centres not stated
• Country/ies: Germany and Italy

Study Duration
2 years

Participants
• Number: 72
• Definition of diagnosis: the 2002 American Thoracic Society/European Respiratory Society International Multidisciplinary Consensus Classification of the Idiopathic Interstitial Pneumonias Statement
• Age: mean 65.9 years (standard deviation 8.7) for low CCL18 group, 69 (8.3) for high CCL18 group
• Gender: 68% male
• Ethnicity: not stated
• Smoking status: 46% ever smokers
• Use of home oxygen therapy: not stated
• Time since diagnosis: mean 18.9 months (standard deviation 20.9) for low CCL18 group, 19.4 (23.1) for high CCL18 group
• Percentage of patients with surgical lung biopsy: not stated
• Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
• Lung function results (% predicted): mean FVC 70.7 (standard deviation 23.4) for low CCL18 group, 66 (22.6) for high CCL18 group; mean DLCO 50.5 (standard deviation 20.2) for low CCL18 group, 44.1 (14.0) for high CCL18 group
| Study Design | Outcomes | Notes |
|--------------|----------|-------|
| Type of Study: randomised controlled trial | Proportion of mortality | Funding source: unclear |
| Trial Design: phase 3, parallel-group, international multicentre (n = 136) | Respiratory-specific mortality | |
| Country/ies: Asia, Australia, Europe, New Zealand, North America, South America | Change in forced vital capacity | |
| Study Duration | Change in diffusing capacity for carbon monoxide | |
| 1 year | Change in 6-minute walk distance | |
| Participants | Notes | |
| Number: 163 (placebo group) | | |
| Definition of diagnosis: the 2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment International Consensus Statement | | |
| Age: mean 65.8 years (standard deviation 7.4) | Proportion of mortality | |
| Gender: 74.2% male | Respiratory-specific mortality | |
| Ethnicity: Caucasian 89.1% | Change in forced vital capacity | |
| Smoking status: 68% ever smoker | Change in diffusing capacity for carbon monoxide | |
| Use of home oxygen therapy: not stated | Change in 6-minute walk distance | |
| Time since diagnosis: mean 1.1 years (standard deviation 1.4) | Notes | |
| Percentage of patients with surgical lung biopsy: 46.8% | | |
| Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated | | |
| Lung function results (% predicted): mean FVC 68.7 (standard deviation 13.1); mean DLCO 42.0 (standard deviation 13.8) | | |
| Resting oxyhaemoglobin saturation: not stated | | |
| 6-minute walk distance: mean 410.4m (standard deviation 118.7) | | |
| Symptom assessment: St George’s Respiratory Questionnaire score: mean 44.5 (standard deviation 21.6); Transition Dyspnoea Index: mean 7.3 (standard deviation 2.4) | | |
| Use of systemic corticosteroid therapy: not stated | | |
| Use of other therapy: not stated | | |

Raghu 2013b [186]

| Study Design | Outcomes | Notes |
|--------------|----------|-------|
| Type of Study: randomised controlled trial | Proportion of mortality | Funding source: industry |
| Trial Design: phase 3, parallel-group, international multicentre (n = 48) | Respiratory-specific mortality | |
| Country/ies: Australia, Canada, France, Germany, Israel, Italy, Slovenia, South Africa, Spain, Sweden, Turkey, United States | Change in forced vital capacity | |
| **Study Duration** | 1 year |
|--------------------|--------|
| **Participants**   |        |
| Number: 59 (placebo group) |
| Definition of diagnosis: the 2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment International Consensus Statement |
| Age: median 64 years (range 49-81) |
| Gender: 62.7% male |
| Ethnicity: not stated |
| Smoking status: 63% ever smokers |
| Use of home oxygen therapy: not stated |
| Time since diagnosis: not stated |
| Percentage of patients with surgical lung biopsy: 100% |
| Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated |
| Lung function results (% predicted): mean FVC 74.8 (standard deviation 14.6); mean DLCO 45.6 (standard deviation 11.2) |
| Resting oxyhaemoglobin saturation: not stated |
| 6-minute walk distance: not stated |
| Symptom assessment: not stated |
| Use of systemic corticosteroid therapy: 25.4% |
| Use of other therapy: N-acetylcysteine 22% |
| **Outcomes** |        |
| Proportion of mortality |
| Change in forced vital capacity |
| Change in diffusing capacity for carbon monoxide |
| **Notes** | Funding source: industry |

Raghu 2014 [122]

| **Study Design** |        |
| Type of Study: retrospective cohort |
| Trial Design: national trial using administrative database (Medicare beneficiaries) |
| Country/ies: United States |
| **Study Duration** | At least 1 year |
| **Participants** |        |
| Number: 12066 |
| Definition of diagnosis: codes using International Classification of Diseases, Ninth Revision, Clinical Modification (ICD-9-CM) |
| Age: mean 79.4 years (standard deviation 7.2) |
| Gender: 46% male |
| Ethnicity: Caucasian 91%, African American 4%, Hispanic 2%, Other 3% |
| Smoking status: not stated |
| Use of home oxygen therapy: not stated |
| Time since diagnosis: not stated |
| Percentage of patients with surgical lung biopsy: not stated |
| Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated |
| Lung function results: not stated |
| Resting oxyhaemoglobin saturation: not stated |
| 6-minute walk distance: not stated |
| Symptom assessment: not stated |
| Use of systemic corticosteroid therapy: not stated |
| Use of other therapy: not stated |
| **Outcomes** | Duration of survival |
| **Notes** | Funding source: industry |
### Raghu 2015 [187]

**Study Design**
- Type of Study: randomised controlled trial
- Trial Design: phase 2, parallel-group international multicentre (number of centres not stated)
- Country/ies: Belgium, Canada, Germany, Netherlands, United States

**Study Duration**
- 72 weeks

**Participants**
- Number: 29 (placebo group)
- Definition of diagnosis: the 2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment International Consensus Statement
- Age: mean 64.5 years (standard deviation 7.26)
- Gender: 79.3% male
- Ethnicity: Caucasian 96.6%, African American 3.4%
- Smoking status: not stated
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: not stated
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: 51.7%
- Lung function results (% predicted): median FEV\(_1\) 70.0 (range 53-89); median FVC 69.0 (range 51-96); median DLCO 38.82 (range 18.0-72.5)
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: median 421.20m (range 157.6-567.0)
- Symptom assessment: total St George’s Respiratory Questionnaire score: median 41.81 (range 12.7-76.2)
- Use of systemic corticosteroid therapy: 13.8%
- Use of other therapy: azathioprine 13.7%, acetylcysteine 31%

**Outcomes**
- Proportion of mortality
- Change in forced vital capacity
- Change in diffusing capacity for carbon monoxide
- Change in health-related quality of life (St George’s Respiratory Questionnaire)

**Notes**
- Funding source: industry

### Raimundo 2016 [123]

**Study Design**
- Type of Study: retrospective cohort
- Trial Design: national using Medicare database
- Country/ies: United States

**Study Duration**
- 1 year

**Participants**
- Number: 13615
- Definition of diagnosis: codes using the International Classification of Diseases, Ninth Revision
- Age: mean age 78.9 years (standard deviation 7.1)
- Gender: 50.3% male
- Ethnicity: not stated
- Smoking status: not stated
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: not stated
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
• Lung function results: not stated
  • Resting oxyhaemoglobin saturation: not stated
  • 6-minute walk distance: not stated
  • Symptom assessment: not stated
  • Use of systemic corticosteroid therapy: not stated
  • Use of other therapy: not stated

Outcomes
• Proportion of mortality
• Duration of survival

Notes
Funding source: unclear

Rajasekaran 2006 [124]

Study Design
• Type of Study: prospective cohort
  • Trial Design: single centre
  • Country/ies: United Kingdom

Study Duration
5 years

Participants
• Number: 18
  • Definition of diagnosis: unclear
  • Age: median 77 years (range 44-88)
  • Gender: 56% male
  • Ethnicity: not stated
  • Smoking status: not stated
  • Use of home oxygen therapy: not stated
  • Time since diagnosis: not stated
  • Percentage of patients with surgical lung biopsy: not stated
  • Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
  • Lung function results (% predicted): median FEV₁ 75 (range 33-109); median FVC 67 (range 47-99); median DLCO 53 (range 17-78)
  • Resting oxyhaemoglobin saturation: not stated
  • 6-minute walk distance: not stated
  • Symptom assessment: not stated
  • Use of systemic corticosteroid therapy: not stated
  • Use of other therapy: not stated

Outcomes
• Proportion of mortality
• Duration of survival

Notes
Funding source: unclear

Rangappa 2009 [125]

Study Design
• Type of Study: retrospective cohort
  • Trial Design: single centre
  • Country/ies: Australia

Study Duration
At least 1 year

Participants
• Number: 24
  • Definition of diagnosis: the 2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment International Consensus Statement
  • Age: mean 66 years (standard deviation 16)
  • Gender: 58% male
  • Ethnicity: not stated
  • Smoking status: not stated
  • Use of home oxygen therapy: 38%
| Study Design | • Type of Study: prospective cohort  
• Trial Design: single centre  
• Country/ies: Scotland |
| Study Duration | 5 years |
| Participants | • Number: 27  
• Definition of diagnosis: the 2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment International Consensus Statement  
• Age: mean 68 years (standard deviation 13) for male, 75 (7) for female  
• Gender: 70% male  
• Ethnicity: not stated  
• Smoking status: 63% ever smokers  
• Use of home oxygen therapy: 41%  
• Time since diagnosis: not stated  
• Percentage of patients with surgical lung biopsy: not stated  
• Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated  
• Lung function results (% predicted): mean DLCO 66 (standard deviation 16) for male, 74 (16) for female  
• Resting oxyhaemoglobin saturation: not stated  
• 6-minute walk distance: not stated  
• Symptom assessment: not stated  
• Use of systemic corticosteroid therapy: 44%  
• Use of other therapy: not stated |
| Outcomes | • Proportion of mortality  
• Duration of survival |
| Notes | Funding source: unclear |

Richards 2012 [127]

| Study Design | • Type of Study: prospective cohort  
• Trial Design: single centre  
• Country/ies: United States |
| Study Duration | At least 1 year |
| Participants | • Number: 140 for derivation group, 101 for validation group  
• Definition of diagnosis: the 2002 American Thoracic Society/European Respiratory Society International Multidisciplinary Consensus Statement |
Classification of the Idiopathic Interstitial Pneumonias Statement

- **Age:** mean 67.2 years (standard deviation 8.3) for derivation group, 68.2 (9.4) for validation group
- **Gender:** 69% male
- **Ethnicity:** Caucasian 97.5%, African American 1%, Native American 0.4%, Oriental 0.4%, unknown 0.4%
- **Smoking status:** 70% ever smoker
- **Use of home oxygen therapy:** not stated
- **Time since diagnosis:** not stated
- **Percentage of patients with surgical lung biopsy:** 52%
- **Percentage of patients with definite usual interstitial pneumonia patterns on HRCT:** not stated
- **Lung function results (% predicted):** mean FVC 62 (standard deviation 19.6) for derivation cohort, 61.4 (18) for validation cohort; mean DLCO 44.8 (standard deviation 17.1) for derivation cohort, 45.4 (19) for validation cohort
- **Resting oxyhaemoglobin saturation:** not stated
- **6-minute walk distance:** not stated
- **Use of systemic corticosteroid therapy:** 23%
- **Use of other therapy:** azathioprine 3%, cyclophosphamide 3%, mycophenolate 0.8%, tacrolimus 0.4%, N-acetylcysteine 0.8%, colchicine 0.4%, interferon-gamma 0.4%

**Outcomes**

- **Duration of survival**
- **Progression-free survival**

**Notes**

Funding source: unclear

Richeldi 2011 [188]

**Study Design**

- **Type of Study:** randomised controlled trial
- **Trial Design:** phase 2, parallel-group, international multicentre (n = 92)
- **Country/ies:** 25 countries

**Study Duration**

1 year

**Participants**

- **Number:** 85 (placebo group)
- **Definition of diagnosis:** the 2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment International Consensus Statement
- **Age:** mean 64.8 years (standard deviation 8.6)
- **Gender:** 74.1% male
- **Ethnicity:** Caucasian 76.5%, Asian 23.5%
- **Smoking status:** not stated
- **Use of home oxygen therapy:** not stated
- **Time since diagnosis:** mean 1.4 years (standard deviation 1.5)
- **Percentage of patients with surgical lung biopsy:** 22.4%
- **Percentage of patients with definite usual interstitial pneumonia patterns on HRCT:** 28.2%
- **Lung function results:** mean FVC 81.7% predicted (standard deviation 17.6); mean DLCO 3.8 mmol/min/kPa (standard deviation 1.1)
- **Resting oxyhaemoglobin saturation:** mean 95.3% (standard deviation 2.2)
- **6-minute walk distance:** not stated
- **Symptom assessment:** total St George’s Respiratory Questionnaire: mean 41.2 (standard deviation 17.9); St George’s Respiratory
Questionnaire-Symptoms score: mean 42.2 (standard deviation 21.6); St George’s Respiratory Questionnaire-Activity score: mean 54.2 (standard deviation 22.2); St George’s Respiratory Questionnaire-Impacts score: mean 33.1 (standard deviation 19.7)

- Use of systemic corticosteroid therapy: 50.6%
- Use of other therapy: not stated

**Outcomes**

- Proportion of mortality
- Respiratory-specific mortality
- Change in forced vital capacity
- Change in health-related quality of life (St George’s Respiratory Questionnaire)

**Notes**

Funding source: industry

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Richeldi 2014 [15]

**Study Design**

- Type of Study: randomised controlled trial
- Trial Design: phase 3, parallel-group, international multicentre (n = 205)
- Country/ies: Asia, Australia, Europe, United States

**Study Duration**

1 year

**Participants**

- Number: 423 (placebo group)
- Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management
- Age: mean 66.9 years (standard deviation 8.2) for INPULSIS-1, 67.1 (7.5) for INPULSIS-2
- Gender: 79% male
- Ethnicity: Caucasian 59%, Asian 30%, unknown 11%
- Smoking status: 71% ever smokers
- Use of home oxygen therapy: not stated
- Time since diagnosis: mean 1.6 years (standard deviation 1.4) for INPULSIS-1, 1.6 (1.3) for INPULSIS-2
- Percentage of patients with surgical lung biopsy: 20%
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results (% predicted): mean FVC 80.5 (standard deviation 17.3) for INPULSIS-1, 78.1 (19) for INPULSIS-2; mean DLCO 47.5 (standard deviation 11.7) for INPULSIS-1, 46.4 (14.8) for INPULSIS-2
- Resting oxyhaemoglobin saturation: mean 95.9% (standard deviation 1.9) for INPULSIS-1, 95.7 (2.1) for INPULSIS-2
- 6-minute walk distance: not stated
- Symptom assessment: Total St George’s Respiratory Questionnaire score: mean 39.8 (standard deviation 18.5) for INPULSIS-1, 39.4 (18.7) for INPULSIS-2
- Use of systemic corticosteroid therapy: 21%
- Use of other therapy: not stated

**Outcomes**

- Proportion of mortality
- Respiratory-specific mortality
- Change in health-related quality of life (St George’s Respiratory Questionnaire)
- Proportion of patients with disease progression

**Notes**

Funding source: industry

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Rogers 2016 [128]
### Study Design
- Type of Study: retrospective cohort
- Trial Design: single centre
- Country/ies: United Kingdom

### Study Duration
At least 1 year

### Participants
- Number: 253
- Definition of diagnosis: ATS/ERS criteria (specific criteria unclear)
- Age: mean 71.4 years (standard deviation 8.3)
- Gender: 74% male
- Ethnicity: not stated
- Smoking status: 70% ever smoker
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: not stated
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results (% predicted): mean FEV₁ 79 (standard deviation 22); mean FVC 82 (standard deviation 19); mean DLCO 45 (standard deviation 15)
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: not stated
- Use of other therapy: not stated

### Outcomes
Duration of survival

### Notes
Funding source: unclear

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**Roig 2010 [129]**

### Study Design
- Type of Study: prospective cohort
- Trial Design: single centre
- Country/ies: Spain

### Study Duration
3 years

### Participants
- Number: 46
- Definition of diagnosis: the 2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment International Consensus Statement
- Age: mean 65 years (range 40-73) for azathioprine group, 63 (40-73) for cyclophosphamide group
- Gender: 63% male
- Ethnicity: not stated
- Smoking status: not stated
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: 56.5%
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results (% predicted): mean FVC 76 (standard deviation 15) for azathioprine group, 76 (10) for cyclophosphamide group; mean DLCO 68 (standard deviation 15) for azathioprine group, 67 (16) for cyclophosphamide group
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: not stated
| Use of systemic corticosteroid therapy: 100% | Use of other therapy: azathioprine 54%, cyclophosphamide 46% |
|-------------------------------------------|---------------------------------------------------------------|
| **Outcomes**                              | Proportion of mortality                                        |
|                                           | Respiratory-specific mortality                                 |
| **Notes**                                 | Funding source: unclear                                        |

### Roskell 2014 [130]

**Study Design**
- Type of Study: prospective cohort for placebo group
- Trial Design: prospective cohort using CPRD-GOLD database
- Country/ies: United Kingdom

**Study Duration**
- At least 1 year

**Participants**
- Number: 193
- Definition of diagnosis: unclear
- Age: not stated
- Gender: not stated
- Ethnicity: not stated
- Smoking status: not stated
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: not stated
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results: not stated
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: not stated
- Use of other therapy: not stated

**Outcomes**
- Duration of survival

**Notes**
- Funding source: unclear

### Rubin 2015 [131]

**Study Design**
- Type of Study: retrospective cohort
- Trial Design: single centre
- Country/ies: Brazil

**Study Duration**
- 1 year

**Participants**
- Number: 44
- Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management
- Age: mean 57 years (range 32-69)
- Gender: 66% male
- Ethnicity: not stated
- Smoking status: not stated
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: not stated
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results (% predicted): mean FEV1 52 (standard deviation 17); mean FVC 50 (standard deviation 18)
| Rudd 2007 [132] | Study Design | Type of Study: prospective cohort  
Trial Design: national  
Country/ies: England, Scotland, Wales |
|---|---|---|
| Study Duration | At least 1 year |
| Participants | Number: 588  
Definition of diagnosis: histological diagnosis of CFA or bilateral interstitial chest radiographic shadowing with bilateral basal inspiratory crackles, and lung function test results compatible with diffuse interstitial fibrosis—that is, a restrictive and/or gas transfer defect.  
Age: mean 67.4 years (standard deviation 10)  
Gender: not stated  
Ethnicity: not stated  
Smoking status: 76% ever smoker  
Use of home oxygen therapy: not stated  
Time since diagnosis: not stated  
Percentage of patients with surgical lung biopsy: 12.4%  
Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated  
Lung function results (% predicted): mean FEV\(_1\) 78.7 (standard deviation 23.7); mean FVC 78.4 (standard deviation 24.8); mean DLCO 49.7 (standard deviation 20.4)  
Resting oxyhaemoglobin saturation: not stated  
6-minute walk distance: not stated  
Symptom assessment: not stated  
Use of systemic corticosteroid therapy: 67%  
Use of other therapy: other drugs (including azathioprine and cyclophosphamide) 14% |
| Outcomes | Proportion of mortality  
Duration of survival |
| Notes | Funding source: unclear |

| Russell 2016 [133] | Study Design | Type of Study: prospective cohort  
Trial Design: single centre study  
Country/ies: United Kingdom |
|---|---|---|
| Study Duration | 3 years |
| Participants | Number: 50  
Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management  
Age: mean 67 years (standard deviation 7.9)  
Gender: 90% male  
Ethnicity: not stated |
| Outcomes | Proportion of mortality  
Duration of survival |
| Notes | Funding source: unclear |
| Smoking status: not stated |
|---------------------------|
| Use of home oxygen therapy: not stated |
| Time since diagnosis: not stated |
| Percentage of patients with surgical lung biopsy: not stated |
| Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated |
| Lung function results (% predicted): mean FEV$_1$ 74.8 (standard deviation 17.0); mean FVC 71.6 (standard deviation 18.3); mean DLCO 39.2 (standard deviation 12.7) |
| Resting oxyhaemoglobin saturation: not stated |
| 6-minute walk distance: not stated |
| Symptom assessment: modified Medical Research Council Dyspnoea score: mean 2.9 (standard deviation 0.9) |
| Use of systemic corticosteroid therapy: not stated |
| Use of other therapy: not stated |

**Outcomes**

- Proportion of mortality
- Duration of survival

**Notes**

- Funding source: public

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**Study Design**

- Type of Study: retrospective cohort
- Trial Design: single centre
- Country/ies: United States

**Study Duration**

At least 1 year

**Participants**

- Number: 307
- Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management
- Age: mean 68.5 years (standard deviation 8.8)
- Gender: 73% male
- Ethnicity: not stated
- Smoking status: not stated
- Use of home oxygen therapy: 18%
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: 41%
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results (% predicted): mean FVC 69.2 (standard deviation 17.9); mean DLCO 45.8 (standard deviation 16.6)
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: not stated
- Use of other therapy: not stated

**Outcomes**

- Duration of survival

**Notes**

- Funding source: public
- Author provided additional data for clarification of study duration and survival/mortality.

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**Sakamoto 2010 [135]**

**Study Design**

- Type of Study: retrospective cohort
### Sato 2016 [136]

| Study Design | Participants | Outcomes |
|--------------|--------------|----------|
| Type of Study: retrospective cohort | Number: 17 | Duration of survival |
| Trial Design: single centre | Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management | Notes |
| Country/ies: Japan | Age: not stated | Funding source: public |
| | Gender: not stated | |
| | Ethnicity: not stated | |
| | Smoking status: not stated | |
| | Use of home oxygen therapy: not stated | |
| | Time since diagnosis: not stated | |
| | Percentage of patients with surgical lung biopsy: not stated | |
| | Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated | |
| | Lung function results: not stated | |
| | Resting oxyhaemoglobin saturation: not stated | |
| | 6-minute walk distance: not stated | |
| | Symptom assessment: not stated | |
| | Use of systemic corticosteroid therapy: not stated | |
| | Use of other therapy: not stated | |

### Notes

Duration of survival

Funding source: public
Saydain 2002 [137]

**Study Design**
- Type of Study: retrospective cohort
- Trial Design: single centre
- Country/ies: United States

**Study Duration** 1 year

**Participants**
- Number: 38
- Definition of diagnosis: the 2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment International Consensus Statement
- Age: not stated
- Gender: 66% male
- Ethnicity: Caucasian 92%, Asian 2.6%, Hispanic 2.6%, Native American Indian 2.6%
- Smoking status: not stated
- Use of home oxygen therapy: 63%
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: not stated
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results (% predicted): mean FVC 56.5 (standard deviation 19.4) for survivors, 59.6 (17.3) for non-survivors; mean DLCO 30.0 (standard deviation 13.1) for survivors, 38.6 (20.3) for non-survivors
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: 53%
- Use of other therapy: not stated

**Outcomes** Proportion of mortality

**Notes** Funding source: public

Schmidt 2011 [138]

**Study Design**
- Type of Study: retrospective cohort
- Trial Design: single centre
- Country/ies: United States

**Study Duration** 1 year

**Participants**
- Number: 321
- Definition of diagnosis: the 2002 American Thoracic Society/European Respiratory Society International Multidisciplinary Consensus Classification of the Idiopathic Interstitial Pneumonias Statement
- Age: mean 63.9 years (standard deviation 9.7)
- Gender: 68% male
- Ethnicity: not stated
- Smoking status: 74% ever smokers
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: not stated
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results (% predicted): mean FEV\(_1\) 79.2 (standard deviation 19.0); mean FVC 67.6 (SD 16.8); mean DLCO 44.5 (SD 16.2)
- Resting oxyhaemoglobin saturation: not stated
| Schupp 2015 [139] | | |
|---|---|---|
| **Study Design** | • Type of Study: prospective cohort  
• Trial Design: single centre  
• Country/ies: Germany |
| **Study Duration** | 2 years |
| **Participants** | • Number: 71  
• Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management  
• Age: mean 68.2 years (standard deviation 8.9) for those with exacerbations, 62.9 (8.0) for those without exacerbations  
• Gender: 83% male  
• Ethnicity: not stated  
• Smoking status: 69% ex-smoker  
• Use of home oxygen therapy: not stated  
• Time since diagnosis (months): mean 18.2 (standard deviation 28.7) for those with exacerbations, 23.4 (19.2) for those without exacerbations  
• Percentage of patients with surgical lung biopsy: not stated  
• Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated  
• Lung function results (% predicted): mean FEV₁ 66.2 (standard deviation 18.1) for those with exacerbations, 54.7 (18.4) for those without exacerbations; mean FVC 67.2 (standard deviation 18.8) for those with exacerbations, 52.3 (20.6) for those without exacerbations; mean DLCO 50.2 (17.9) for those with exacerbations, 28.3 (2.4) for those without exacerbations  
• Resting oxyhaemoglobin saturation: not stated  
• 6-minute walk distance: not stated  
• Symptom assessment: not stated  
• Use of systemic corticosteroid therapy: not stated  
• Use of other therapy: not stated |
| **Outcomes** | • Proportion of mortality |
| **Notes** | Funding source: public |

| Selman 1998 [140] | | |
|---|---|---|
| **Study Design** | • Type of Study: prospective cohort  
• Trial Design: single centre  
• Country/ies: Mexico |
| **Study Duration** | At least 1 year |
| **Participants** | • Number: 56  
• Definition of diagnosis: progressive dyspnoea, diffuse reticulonodular infiltrates on CXR, bibasilar crackles, digital clubbing, decreased FVC and PaO2 and no evidence of systemic disease or environmental exposure  
• Age: mean 55 years (standard deviation 10) for prednisolone group, 55 |
| **Outcomes** | | |
| **Notes** | Proportion of mortality  
Funding source: public |
| Outcomes                                      | Proportion of mortality                                                                 |
|----------------------------------------------|----------------------------------------------------------------------------------------|
| Notes                                        | Funding source: unclear                                                                  |

Selman 2007 [141]

| Study Design                                  | Type of Study: retrospective cohort                                                     |
|----------------------------------------------|----------------------------------------------------------------------------------------|
|                                              | Trial Design: single centre                                                              |
|                                              | Country/ies: Mexico                                                                     |

| Study Duration                               | 1 year                                                                                 |
|----------------------------------------------|----------------------------------------------------------------------------------------|

| Participants                                  | Number: 114                                                                            |
|----------------------------------------------|----------------------------------------------------------------------------------------|
|                                              | Definition of diagnosis: the 2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment International Consensus Statement |
|                                              | Age: mean 64.1 years (standard deviation 12.1) for rapid progressors, 63.6 (8.8) for slow progressors |
|                                              | Gender: 71% male                                                                       |
|                                              | Ethnicity: not stated                                                                   |
|                                              | Smoking status: 58% ever smokers                                                       |
|                                              | Use of home oxygen therapy: not stated                                                 |
|                                              | Time since diagnosis: not stated                                                       |
|                                              | Percentage of patients with surgical lung biopsy: 31%                                  |
|                                              | Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated |
|                                              | Lung function results (% predicted): mean FVC 58.4 (standard deviation 21.6) for rapid progressors, 61.5 (18.1) for slow progressors |
|                                              | Resting oxyhaemoglobin saturation: mean 85.6% (standard deviation 7.6) for rapid progressors, 82.0% (standard deviation 10.4) for slow progressors |
|                                              | 6-minute walk distance: not stated                                                     |
|                                              | Symptom assessment: not stated                                                         |
Use of systemic corticosteroid therapy: not stated
Use of other therapy: not stated

**Outcomes**

Duration of survival

**Notes**

Funding source: public

### Serrano-Mollar 2016 [142]

**Study Design**
- Type of Study: prospective cohort
- Trial Design: national (number of centres not specified)
- Country/ies: Spain

**Study Duration**
1 year

**Participants**
- Number: 16
- Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management
- Age: mean 61.3 years (range 51-73)
- Gender: 81% male
- Ethnicity: not stated
- Smoking status: 88% ever smokers
- Use of home oxygen therapy: not stated
- Time since diagnosis: median 21 months (range 6-36)
- Percentage of patients with surgical lung biopsy: not stated
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results (% predicted): median FVC 67 (range 50-105); median DLCO 48.5 (range 35-59)
- Resting oxyhaemoglobin saturation: median 95.5% (range 92-98)
- 6-minute walk distance: median 552.5m (range 342-690)
- Symptom assessment: Basal Dyspnoea Index: median 9 (range 4-12); Leicester Cough Questionnaire: median 18.9 (range 5.5-21)
- Use of systemic corticosteroid therapy: not stated
- Use of other therapy: not stated

**Outcomes**

Duration of survival

**Notes**

Funding source: public

### Shafiq 2010 [143]

**Study Design**
- Type of Study: retrospective cohort
- Trial Design: single centre
- Country/ies: United States

**Study Duration**
1 year

**Participants**
- Number: 27
- Definition of diagnosis: unclear
- Age: not stated
- Gender: not stated
- Ethnicity: not stated
- Smoking status: not stated
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: not stated
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results: FVC 2.7 +/- 0.2 L; DLCO 2.4 +/- 0.8 ml/min/mmHg
• Resting oxyhaemoglobin saturation: not stated
• 6-minute walk distance: not stated
• Symptom assessment: not stated
• Use of systemic corticosteroid therapy: not stated
• Use of other therapy: not stated

**Outcomes**
Change in diffusing capacity for carbon monoxide

**Notes**
Funding source: unclear

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**Sherbini 2014** [144]

**Study Design**
- Type of Study: retrospective cohort
- Trial Design: national multicentre (n = 2)
- Country/ies: Saudi Arabia

**Study Duration**
Greater than 1 year

**Participants**
- Number: 134
- Definition of diagnosis: the 2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment International Consensus Statement
- Age: mean 64 years (standard deviation 13)
- Gender: 44% male
- Ethnicity: not stated
- Smoking status: 36% ever smokers
- Use of home oxygen therapy: 53%
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: not stated
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results: mean FEV₁ 56 (standard deviation 15); mean FVC 53 (standard deviation 13); mean DLCO 57 (standard deviation 15)
- Resting oxyhaemoglobin saturation: mean 92% (standard deviation 7)
- 6-minute walk distance: mean 338m (standard deviation 64)
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: 84%
- Use of other therapy: azathioprine 69%, colchicine 20%, N-acetylcysteine 11%, omeprozole 67%

**Outcomes**
Proportion of mortality

**Notes**
Funding source: unclear

---

**Shin 2008** [145]

**Study Design**
- Type of Study: retrospective cohort
- Trial Design: single centre
- Country/ies: Korea

**Study Duration**
At least 1 year (mean 45 months)

**Participants**
- Number: 79
- Definition of diagnosis: histological usual interstitial pneumonia, excluded connective tissue disease related-ILD and hypersensitivity pneumonitis; or the 2002 American Thoracic Society/European Respiratory Society International Multidisciplinary Consensus Classification of the Idiopathic Interstitial Pneumonias Statement with ≥ 10% honeycombing
- Age: mean 63 years (standard deviation 7.4)
- Gender: 76% male
- Ethnicity: not stated
| Study Design | Participants |
|--------------|--------------|
| • Smoking status: 66% ever smokers<br>• Use of home oxygen therapy: not stated<br>• Time since diagnosis: not stated<br>• Percentage of patients with surgical lung biopsy: 68%<br>• Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: 32%<br>• Lung function results (% predicted): mean FVC 74 (standard deviation 15); mean DLCO 66 (standard deviation 28)<br>• Resting oxyhaemoglobin saturation: not stated<br>• 6-minute walk distance: not stated<br>• Symptom assessment: not stated<br>• Use of systemic corticosteroid therapy: not stated<br>• Use of other therapy: immunosuppressive therapies 70% (specific therapy not stated) | • Number: 39<br>• Definition of diagnosis: the 2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment International Consensus Statement<br>• Age: median 58 years (interquartile range 56-66) for survivors, 66 (interquartile range: 60-73) for non-survivors<br>• Gender: 74% male<br>• Ethnicity: not stated<br>• Smoking status: 64% ever smoker<br>• Use of home oxygen therapy: not stated<br>• Time since diagnosis: not stated<br>• Percentage of patients with surgical lung biopsy: not stated<br>• Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated<br>• Lung function results (% predicted): median FEV₁ 79.3 (interquartile range 76.1-87.6) for survivors, 82.5 (74.0-94.0) for non-survivors; median FVC 70 (interquartile range 63-79) for survivors, 72 (interquartile range 57-89) for non-survivors; median DLCO 51.5 (interquartile range 42.6-68.3) for survivors, 34.0 (interquartile range 26.3-38.8) for non-survivors<br>• Resting oxyhaemoglobin saturation: not stated<br>• 6-minute walk distance: not stated<br>• Symptom assessment: not stated<br>• Use of systemic corticosteroid therapy: not stated<br>• Use of other therapy: not stated |

**Outcomes**

Proportion of mortality

**Notes**

Funding source: public

**Shinoda 2009 [146]**

**Study Design**

- Type of Study: retrospective cohort
- Trial Design: single centre
- Country/ies: Japan

**Study Duration**

5 years

**Participants**

- Number: 39
- Definition of diagnosis: the 2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment International Consensus Statement
- Age: median 58 years (interquartile range 56-66) for survivors, 66 (interquartile range: 60-73) for non-survivors
- Gender: 74% male
- Ethnicity: not stated
- Smoking status: 64% ever smoker
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: not stated
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results (% predicted): median FEV₁ 79.3 (interquartile range 76.1-87.6) for survivors, 82.5 (74.0-94.0) for non-survivors; median FVC 70 (interquartile range 63-79) for survivors, 72 (interquartile range 57-89) for non-survivors; median DLCO 51.5 (interquartile range 42.6-68.3) for survivors, 34.0 (interquartile range 26.3-38.8) for non-survivors
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: not stated
- Use of other therapy: not stated

**Outcomes**

Proportion of mortality

**Notes**

Funding source: Unclear

**Shitrit 2009 [147]**

**Study Design**

- Type of Study: prospective cohort
### Study Design
- Type of Study: retrospective cohort
- Trial Design: national using the Organ Procurement and Transplantation Network (OPTN) database
- Country/ies: United States

### Study Duration
- 1 year

### Participants
- Number: 1339
- Definition of diagnosis: unclear
- Age: not stated
- Gender: 67% male
- Ethnicity: Caucasian 79%, Hispanic 11%, African American 7%, other 3%
- Smoking status: not stated
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: not stated
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results: not stated
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: 765 - 960 feet
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: not stated
- Use of other therapy: not stated

### Outcomes
- Proportion of mortality

### Notes
- Funding source: unclear
### Siemienowicz 2015 [149]

#### Study Design
- Type of Study: retrospective cohort
- Trial Design: single centre
- Country/ies: Australia

#### Study Duration
- 5 years

#### Participants
- Number: 20
- Definition of diagnosis: based on medical records
- Age: mean 75.3 years (standard deviation 8.8)
- Gender: 65% male
- Ethnicity: not stated
- Smoking status: 65% ever smoker
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: 15%
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results: not stated
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: not stated
- Use of other therapy: not stated

#### Outcomes
- Proportion of mortality

#### Notes
- Funding source: unclear
- Author provided additional data for clarification of study duration and survival/mortality.

### Soares Pires 2013 [150]

#### Study Design
- Type of Study: retrospective cohort
- Trial Design: single centre
- Country/ies: Portugal

#### Study Duration
- 4 years

#### Participants
- Number: 81
- Definition of diagnosis: the 2002 American Thoracic Society/European Respiratory Society International Multidisciplinary Consensus Classification of the Idiopathic Interstitial Pneumonias Statement
- Age: mean 63.8 years (standard deviation 10.2)
- Gender: 69.1% male
- Ethnicity: not stated
- Smoking status: 51.9% ever smoker
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: 30.9%
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results: mean FEV$_1$ 80.8 (standard deviation 19.2); mean FVC 74.8 (standard deviation 20.2); mean DLCO 45.8 (standard deviation 16.4)
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: mean 369.6m (standard deviation 149.4)
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: 86.4%
Soares 2015 [151]

**Study Design**
- Type of Study: retrospective cohort
- Trial Design: multicentre national (n =3)
- Country/ies: Brazil

**Study Duration**
At least 1 year

**Participants**
- Number: 120
- Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management
- Age: mean 68.6 years (standard deviation 7.9)
- Gender: 70% male
- Ethnicity: not stated
- Smoking status: 61.2% ever smoker
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: 30.8%
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results (% predicted): mean FEV1 79.1 (standard deviation 15.1); mean FVC 75.2 (standard deviation 15.3); mean DLCO 47.1 (standard deviation 13.5)
- Resting oxyhaemoglobin saturation: mean 94.4% (standard deviation 2.2)
- 6-minute walk distance: not stated
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: not stated
- Use of other therapy: not stated

**Outcomes**
- Proportion of mortality
- Duration of survival

**Notes**
- Funding source: unclear
- Same cohort as above but different outcomes have been measured in this study

Sobiecka 2013 [152]

**Study Design**
- Type of Study: retrospective cohort
- Trial Design: single centre
- Country/ies: Poland

**Study Duration**
At least 1 year

**Participants**
- Number: 56
- Definition of diagnosis: unclear
- Age: mean 62 +/- 10 years
- Gender: 57% male
- Ethnicity: not stated
- Smoking status: 55% ever smoker
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: not stated
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results: not stated
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: not stated
- Use of other therapy: not stated

### Outcomes
- Duration of survival

### Notes
- Funding source: unclear

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**Song 2011 [153]**

| Study Design |
|--------------|
| - Type of Study: retrospective cohort  
- Trial Design: single centre  
- Country/ies: Korea |

| Study Duration |
|---------------|
| At least 1 year |

| Participants |
|--------------|
| - Number: 461  
- Definition of diagnosis: the 2002 American Thoracic Society/European Respiratory Society International Multidisciplinary Consensus Classification of the Idiopathic Interstitial Pneumonias Statement  
- Age: mean 63.4 years  
- Gender: 77.7% male  
- Ethnicity: not stated  
- Smoking status: not stated  
- Use of home oxygen therapy: not stated  
- Time since diagnosis: not stated  
- Percentage of patients with surgical lung biopsy: not stated  
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated  
- Lung function results: mean FEV₁ 88.5 (standard deviation 18) for non-rapid deterioration group, 86 (18) for acute exacerbation group, 89 (17.5) for infection group; mean FVC 77.6 (standard deviation 17) for non-rapid deterioration group, 72 (15.7) for acute exacerbation group, 75.5 (18.5) for infection group; mean DLCO 66.4 (standard deviation 19) for non-rapid deterioration group, 62.2 (19.3) for acute exacerbation group, 61.2 (18) for infection group  
- Resting oxyhaemoglobin saturation: not stated  
- 6-minute walk distance: not stated  
- Symptom assessment: not stated  
- Use of systemic corticosteroid therapy: 56%  
- Use of other therapy: not stated |

| Outcomes |
|----------|
| Proportion of mortality |

| Notes |
|-------|
| Funding source: unclear |

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**Song 2014 [154]**

| Study Design |
|--------------|
| - Type of Study: retrospective cohort  
- Trial Design: single centre  
- Country/ies: Korea |

| Study Duration |
|---------------|
| 1 year |

| Participants |
|--------------|
| - Number: 43  
- Definition of diagnosis: histologically usual interstitial pneumonia, |
| Study Design | Stack 1972 [155] |
|--------------|------------------|
| Type of Study: | retrospective cohort |
| Trial Design:  | single centre |
| Country/ies:   | United Kingdom |

### Study Duration

- 1 year

### Participants

- Number: 96
- Definition of diagnosis: progressive non-episodic dyspnoea without wheeze, bilateral crepitations on auscultation of the chest, and diffuse bilateral pulmonary opacities on chest radiograph
- Age: 51% male
- Gender: not stated
- Ethnicity: not stated
- Smoking status: not stated
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: 50%
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results: not stated
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: not stated
- Use of other therapy: not stated

### Outcomes

- Proportion of mortality

### Notes

- Funding source: unclear

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| Study Design | Strand 2014 [156] |
|--------------|--------------------|
| Type of Study: | prospective cohort |
| Trial Design:  | single centre |
| Country/ies:   | United States |

### Study Duration

- 1 year

### Participants

- Number: 96
- Definition of diagnosis: progressive non-episodic dyspnoea without wheeze, bilateral crepitations on auscultation of the chest, and diffuse bilateral pulmonary opacities on chest radiograph
- Age: 51% male
- Gender: not stated
- Ethnicity: not stated
- Smoking status: not stated
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: 50%
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results: not stated
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: not stated
- Use of other therapy: not stated

### Outcomes

- Duration of survival

### Notes

- Funding source: unclear
| Study Duration | At least 1 year |
|----------------|----------------|
| **Participants** |  |
| • Number: 321 |  |
| • Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management |  |
| • Age: mean 66.1 years (standard deviation 9.1) |  |
| • Gender: 75% male |  |
| • Ethnicity: Caucasian 97.8%, Asian 1.3%, African American 0.3%, Other 0.6% |  |
| • Smoking status: not stated |  |
| • Use of home oxygen therapy: not stated |  |
| • Time since diagnosis: not stated |  |
| • Percentage of patients with surgical lung biopsy: 321 (100%) |  |
| • Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated |  |
| • Lung function results (% predicted): mean FVC 71.4 (standard deviation 17.4), mean DLCO 52.3 (SD 18.7) |  |
| • Resting oxyhaemoglobin saturation: not stated |  |
| • 6-minute walk distance: not stated |  |
| • Symptom assessment: not stated |  |
| • Use of systemic corticosteroid therapy: not stated |  |
| • Use of other therapy: not stated |  |
| **Outcomes** | Duration of survival |
| **Notes** | Funding source: public |

**Strongman 2018 [157]**

| Study Design |  |
| • Type of Study: retrospective cohort |  |
| • Trial Design: national using the Clinical Practice Research Datalink GOLD dataset |  |
| • Country/ies: UK |  |
| **Study Duration** | 1 year or greater |
| **Participants** |  |
| • Number: 1389 |  |
| • Definition of diagnosis: using specific disease Read codes |  |
| • Age: not stated |  |
| • Gender: not stated |  |
| • Ethnicity: not stated |  |
| • Smoking status: not stated |  |
| • Use of home oxygen therapy: not stated |  |
| • Time since diagnosis: not stated |  |
| • Percentage of patients with surgical lung biopsy: not stated |  |
| • Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated |  |
| • Lung function results: not stated |  |
| • Resting oxyhaemoglobin saturation: not stated |  |
| • 6-minute walk distance: not stated |  |
| • Symptom assessment: not stated |  |
| • Use of systemic corticosteroid therapy: not stated |  |
| • Use of other therapy: not stated |  |
| **Outcomes** | Proportion of mortality |
| **Notes** | Funding source: industry |
| • Author provided additional data for clarification of study duration and survival/mortality. |  |

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**Su 2011 [158]**

| **Study Design** |  |
|------------------|--|
| Type of Study: retrospective cohort |  |
| Trial Design: single centre |  |
| Country/ies: United States |  |

| **Study Duration** | At least 1 year |
|--------------------|-----------------|

| **Participants** |  |
|------------------|--|
| Number: 148 |  |
| Definition of diagnosis: the 2002 American Thoracic Society/European Respiratory Society International Multidisciplinary Consensus Classification of the Idiopathic Interstitial Pneumonias Statement |  |
| Age: mean 68.6 years (standard deviation 12.1) |  |
| Gender: 58% male |  |
| Ethnicity: Caucasian 69% |  |
| Smoking status: 68% ever smokers |  |
| Use of home oxygen therapy: not stated |  |
| Time since diagnosis: mean 1.7 years (standard deviation 4) |  |
| Percentage of patients with surgical lung biopsy: 68% |  |
| Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated |  |
| Lung function results (% predicted): mean FEV$$_1$$ 75.9 (standard deviation 23.3); mean FVC 67.3 (standard deviation 20.2); mean DLCO 44.9 (standard deviation 18.7) |  |
| Resting oxyhaemoglobin saturation: not stated |  |
| 6-minute walk distance: not stated |  |
| Symptom assessment: not stated |  |
| Use of systemic corticosteroid therapy: 48.7% |  |
| Use of other therapy: cyclophosphamide 8.1%, azathioprine 37.8%, methotrexate 1.4%, mycophenolate 14.9%, hydroxychloroquine 6.1% |  |

| **Outcomes** | Proportion of mortality |
|--------------|-------------------------|

| **Notes** |  |
|------------|-------------------------|
| Funding source: unclear |  |
| Author provided additional data for clarification of study duration and survival/mortality. |  |

**Takahashi 2000 [159]**

| **Study Design** |  |
|------------------|--|
| Type of Study: retrospective cohort |  |
| Trial Design: single centre |  |
| Country/ies: Japan |  |

| **Study Duration** | At least 1 year |
|--------------------|-----------------|

| **Participants** |  |
|------------------|--|
| Number: 52 |  |
| Definition of diagnosis: either evidence of varying degrees of interstitial fibrosis and alveolitis or evidence of diffuse parenchymal infiltrates on chest radiography |  |
| Age: mean 62.5 years (standard deviation 7.5) |  |
| Gender: 83% male |  |
| Ethnicity: not stated |  |
| Smoking status: 87% ever smoker |  |
| Use of home oxygen therapy: not stated |  |
| Time since diagnosis: not stated |  |
| Percentage of patients with surgical lung biopsy: not stated |  |
| Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated |  |
| Study Design | Taniguchi 2010 [189] |
|--------------|----------------------|
| Type of Study: randomised controlled trial |
| Trial Design: phase 3, parallel-group, national multicentre (n = 73) |
| Country/ies: Japan |

| Study Duration | 1 year |
|----------------|--------|

| Participants | Taniguchi 2010 [189] |
|--------------|----------------------|
| Number: 104 (placebo group) |
| Definition of diagnosis: the 2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment International Consensus Statement |
| Age: mean 64.7 years (standard deviation 7.3) |
| Gender: 77.9% male |
| Ethnicity: not stated |
| Smoking status: 80% ever smokers |
| Use of home oxygen therapy: not stated |
| Time since diagnosis: not stated |
| Percentage of patients with surgical lung biopsy: 26.9% |
| Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated |
| Lung function results (% predicted): mean FVC 79.1 (standard deviation 17.4); mean 55.2 (SD 18.2) |
| Resting oxyhaemoglobin saturation: not stated |
| 6-minute walk distance: not stated |
| Symptom assessment: not stated |
| Use of systemic corticosteroid therapy: 4.8% |
| Use of other therapy: not stated |

| Outcomes | Proportion of mortality |
|----------|-------------------------|
| Notes | Funding source: public |

| Study Design | ten Klooster 2015 [160] |
|--------------|------------------------|
| Type of Study: prospective cohort |
| Trial Design: unclear |
| Country/ies: Netherlands |

| Study Duration | 4 years |
|----------------|---------|

| Participants | ten Klooster 2015 [160] |
|--------------|------------------------|
| Number: 169 |
| Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management |
| Age: mean 59.4 years (standard deviation 12.9) for initial group, 62.9 (8.7) for duplication group |
| Gender: 83% male |
| Ethnicity: not stated |
| Smoking status: 66% ever smokers, 0.08% unknown |

| Outcomes | Proportion of patients with disease progression |
|----------|------------------------------------------------|
| Notes | Funding source: public |
Use of home oxygen therapy: not stated
Time since diagnosis: not stated
Percentage of patients with surgical lung biopsy: not stated
Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
Lung function results (% predicted): mean FVC 72.6 (standard deviation 23.5) for initial group, 82.8 (24.3) for duplication group; mean DLCO 45.1 (standard deviation 16.3) for initial group, 46.6 (15.1) for duplication group
Resting oxyhaemoglobin saturation: not stated
6-minute walk distance: not stated
Symptom assessment: not stated
Use of systemic corticosteroid therapy: not stated
Use of other therapy: not stated

Outcomes
- Proportion of mortality
- Duration of survival

Notes
- Funding source: unclear

Toello 2014 [162]

| Study Design | Type of Study: retrospective cohort
| | Trial Design: single centre
| | Country/ies: United States

| Study Duration | At least 1 year

| Participants | Number: 133
| | Definition of diagnosis: multidisciplinary diagnosis of IPF, with surgical
biopsy showing usual interstitial pneumonia pattern
- Age: mean 63.6 years (standard deviation 8.1)
- Gender: 63.2% male
- Ethnicity: not stated
- Smoking status: 62.1% ever smokers
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: 100%
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: 30.8%
- Lung function results (% predicted): mean FVC 67.3 (standard deviation 15.8); mean DLCO 48.9 (standard deviation 15.9)
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: not stated
- Use of other therapy: not stated

**Outcomes**
Duration of survival

**Notes**
Funding source: public

| Tomioka 2007 [163] |
|-------------------|
| **Study Design** |
| - Type of Study: prospective cohort |
| - Trial Design: single centre |
| - Country/ies: Japan |
| **Study Duration** |
| 1 year |
| **Participants** |
| - Number: 46 |
| - Definition of diagnosis: the 2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment International Consensus Statement |
| - Age: mean 69.9 years (standard deviation 5.8) |
| - Gender: 70% male |
| - Ethnicity: not stated |
| - Smoking status: 70% ever smokers |
| - Use of home oxygen therapy: 15% |
| - Time since diagnosis: not stated |
| - Percentage of patients with surgical lung biopsy: not stated |
| - Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated |
| - Lung function results (% predicted): mean FVC 71.0 (standard deviation 17.5); mean DLCO 58.3 (standard deviation 18.2) |
| - Resting oxyhaemoglobin saturation: not stated |
| - 6-minute walk distance: 395m (standard deviation 105) |
| - Symptom assessment: not stated |
| - Use of systemic corticosteroid therapy: 4% |
| - Use of other therapy: not stated |
| **Outcomes** |
| Proportion of mortality |
| **Notes** |
| Funding source: unclear |

| Tryfon 2009 [164] |
|-------------------|
| **Study Design** |
| - Type of Study: prospective cohort |
| - Trial Design: single centre |
### Tzouvelekis 2013 [165]

| **Study Design** |  |
|---|---|
| **Type of Study:** | prospective cohort |
| **Trial Design:** | single centre |
| **Country/ies:** | Greece |

| **Study Duration** | 1 year |

| **Participants** |  |
|---|---|
| **Number:** | 14 |
| **Definition of diagnosis:** | the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management |
| **Age:** | mean 64.4 years (standard deviation 7) |
| **Gender:** | 86% male |
| **Ethnicity:** | not stated |
| **Smoking status:** | 100% ever smokers |
| **Use of home oxygen therapy:** | not stated |
| **Time since diagnosis:** | not stated |
| **Percentage of patients with surgical lung biopsy:** | 43% |
| **Percentage of patients with definite usual interstitial pneumonia patterns on HRCT:** | 100% |
| **Lung function results (% predicted):** | mean FVC 71.2 (standard deviation 15.2); mean DLCO 48.4 (standard deviation 11.1) |
| **Resting oxyhaemoglobin saturation:** | not stated |
| **6-minute walk distance:** | mean 472.1m (standard deviation 55.2) |
| **Symptom assessment:** | modified Medical Research Council Dyspnoea score: mean 2.1 (standard deviation 0.6); St George’s Respiratory Questionnaire score: mean 35.1 (standard deviation 6.8) |
| **Use of systemic corticosteroid therapy:** | 36% |
| **Use of other therapy:** | N-acetylcysteine 36% |

| **Outcomes** | Proportion of mortality |

| **Notes** | Funding source: industry |
### Tzouvelekis 2016 [166]

| **Study Design** |  |
|------------------|------------------|
| Type of Study:    | prospective cohort |
| Trial Design:     | single centre     |
| Country/ies:      | United States     |

| **Study Duration** | At least 1 year |

| **Participants** |  |
|------------------|------------------|
| Number:          | 97               |
| Definition of diagnosis: | the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management |
| Age:             | mean age 70 years (standard deviation 8) |
| Gender:          | 78.4% male       |
| Ethnicity:       | Caucasian 87.6%   |
| Smoking status:  | 72% ever smokers |
| Use of home oxygen therapy: | 50.5% |
| Time since diagnosis: | not stated |
| Percentage of patients with surgical lung biopsy: | 23.7% |
| Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: | not stated |
| Lung function results: | not stated |
| Resting oxyhaemoglobin saturation: | not stated |
| 6-minute walk distance: | not stated |
| Symptom assessment: | not stated |
| Use of systemic corticosteroid therapy: | not stated |
| Use of other therapy: | not stated |

| **Outcomes** | Proportion of mortality |

| **Notes** |  |
|-----------|--------------------------|
| Funding source: | public |
| Author provided additional data for clarification of study duration and survival/mortality. |

### Umeda 2015 [167]

| **Study Design** |  |
|------------------|------------------|
| Type of Study:    | prospective cohort |
| Trial Design:     | single centre     |
| Country/ies:      | Japan             |

| **Study Duration** | At least 1 year |

| **Participants** |  |
|------------------|------------------|
| Number:          | 50               |
| Definition of diagnosis: | the 2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment International Consensus Statement |
| Age:             | mean 70.4 years (standard deviation 9.0) |
| Gender:          | 84% male         |
| Ethnicity:       | not stated       |
| Smoking status:  | 74% ever smokers |
| Use of home oxygen therapy: | not stated |
| Time since diagnosis: | not stated |
| Percentage of patients with surgical lung biopsy: | not stated |
| Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: | not stated |
| Lung function results (% predicted): | mean FVC 84.1 (standard deviation 24.3); mean DLCO 56.6 (standard deviation 18) |
| Resting oxyhaemoglobin saturation: | not stated |
| 6-minute walk distance: | mean 370m (standard deviation 123) |
|                  | **Symptom assessment:** not stated |
|------------------|----------------------------------|
|                  | **Use of systemic corticosteroid therapy:** not stated |
|                  | **Use of other therapy:** not stated |

**Outcomes**
- Proportion of mortality
- Progression-free survival

**Notes**
- Funding source: public

### Vysehradsky 2002 [168]

| **Study Design** |                                                          |
|------------------|----------------------------------------------------------|
|                  | **Type of Study:** retrospective cohort                   |
|                  | **Trial Design:** single centre                           |
|                  | **Country/ies:** Slovakia                                 |

| **Study Duration** | 1 to 10 years |

| **Participants** |                                                          |
|------------------|----------------------------------------------------------|
|                  | **Number:** 34                                           |
|                  | **Definition of diagnosis:** lung biopsy or the British Thoracic Society algorithm based on clinical assessment and investigations |
|                  | **Age:** mean 43.5 years (standard deviation 13.7)        |
|                  | **Gender:** 44% male                                      |
|                  | **Ethnicity:** not stated                                  |
|                  | **Smoking status:** not stated                             |
|                  | **Use of home oxygen therapy:** not stated                |
|                  | **Time since diagnosis:** mean 32 months (standard deviation 33.6) |
|                  | **Percentage of patients with surgical lung biopsy:** 47% |
|                  | **Percentage of patients with definite usual interstitial pneumonia patterns on HRCT:** not stated |
|                  | **Lung function results:** (% predicted): mean FVC 80.9% (standard deviation 19.6); mean DLCO 48.8 (standard deviation 13.1) |
|                  | **Resting oxyhaemoglobin saturation:** not stated         |
|                  | **6-minute walk distance:** not stated                    |
|                  | **Symptom assessment:** not stated                         |
|                  | **Use of systemic corticosteroid therapy:** 94%            |
|                  | **Use of other therapy:** azathioprine 19%, cyclophosphamide 38%, colchicine 41%, D-penicilliamine 6% |

| **Outcomes** |                                                          |
|--------------|----------------------------------------------------------|
|              | **Change in forced vital capacity:** (data available in proportion of patients who deteriorated) |
|              | **Change in diffusing capacity for carbon monoxide:** (data available in proportion of patients who deteriorated) |

| **Notes** | Funding source: unclear |

### Wallaert 2011 [169]

| **Study Design** |                                                          |
|------------------|----------------------------------------------------------|
|                  | **Type of Study:** retrospective cohort                   |
|                  | **Trial Design:** national multicentre (n = 2)            |
|                  | **Country/ies:** France                                   |

| **Study Duration** | 3 years |

| **Participants** |                                                          |
|------------------|----------------------------------------------------------|
|                  | **Number:** 63                                           |
|                  | **Definition of diagnosis:** the 2002 American Thoracic Society/European Respiratory Society International Multidisciplinary Consensus Classification of the Idiopathic Interstitial Pneumonias Statement |
|                  | **Age:** mean 65 +/- 8 years for group 1, 63 +/- 6.7 for group 2 |
|                  | **Gender:** 81% male                                      |
|                  | **Ethnicity:** not stated                                  |
|                  | **Smoking status:** not stated                             |
|                  | **Use of home oxygen therapy:** not stated                |
Wilkie 2012 [170]

**Study Design**
- Type of Study: retrospective cohort
- Trial Design: single centre
- Country/ies: United Kingdom

**Study Duration**
- 3 years

**Participants**
- Number: 88
- Definition of diagnosis: unclear
- Age: median 69 years (interquartile range 63-76)
- Gender: 57.5% male
- Ethnicity: not stated
- Smoking status: not stated
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: not stated
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results: not stated
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: not stated
- Use of other therapy: not stated

**Outcomes**
- Proportion of mortality

**Notes**
- Funding source: unclear

Wright 1981 [171]

**Study Design**
- Type of Study: retrospective cohort
- Trial Design: national multicentre (n =2)
- Country/ies: United Kingdom

**Study Duration**
- At least 2 years

**Participants**
- Number: 62
- Definition of diagnosis: trephine lung biopsy with changes of cryptogenic fibrosing alveolitis; patients with history of exposure to industrial or other dusts likely to produce parenchymal lung disease, or a history of taking drugs known to causes such disease, or clinical evidence of
rheumatoid arthritis or collagen vascular disorder were excluded

- Age: mean 57+/-11 years
- Gender: 76% male
- Ethnicity: not stated
- Smoking status: not stated
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: 100%
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results (% predicted): mean FVC 77 +/- 25; mean DLCO 44 +/- 15
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: 65%
- Use of other therapy: not stated

### Outcomes
Proportion of mortality

### Notes
Funding source: unclear

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**Yokoyama 2006 [172]**

| **Study Design** | Type of Study: retrospective cohort |
|------------------|-------------------------------------|
|                  | Trial Design: national multicentre (n = 7) |
|                  | Country/ies: Japan |

| **Study Duration** | 3 years |
|--------------------|---------|

| **Participants** | Number: 27 |
|------------------|------------|
|                  | Definition of diagnosis: the 2000 American Thoracic Society/European Respiratory Society Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment International Consensus Statement |
|                  | Age: mean 59 +/- 10 years |
|                  | Gender: 70% male |
|                  | Ethnicity: not stated |
|                  | Smoking status: not stated |
|                  | Use of home oxygen therapy: not stated |
|                  | Time since diagnosis: not stated |
|                  | Percentage of patients with surgical lung biopsy: 59% |
|                  | Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: 41% |
|                  | Lung function results: not stated |
|                  | Resting oxyhaemoglobin saturation: not stated |
|                  | 6-minute walk distance: not stated |
|                  | Symptom assessment: not stated |
|                  | Use of systemic corticosteroid therapy: not stated |
|                  | Use of other therapy: not stated |

| **Outcomes** | Proportion of mortality |
|--------------|-------------------------|

### Notes
Funding source: Public

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**Yu 2014 [173]**

| **Study Design** | Type of Study: retrospective cohort |
|------------------|-------------------------------------|
|                  | Trial Design: national using the Department of Defense Military Health System database |
|                  | Country/ies: United States |

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223
### Study Duration
At least 1 year

### Participants
- Number: 67
- Definition of diagnosis: International Classification of Diseases, Ninth Revision
- Age: mean 66 years
- Gender: 57% male
- Ethnicity: not stated
- Smoking status: not stated
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: not stated
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results: not stated
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: not stated
- Use of other therapy: not stated

### Outcomes
Proportion of mortality

### Notes
Funding source: unclear

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**Zhang 2016 [174]**

### Study Design
- Type of Study: retrospective cohort
- Trial Design: single centre
- Country/ies: China

### Study Duration
5 years

### Participants
- Number: 105
- Definition of diagnosis: the 2011 Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management
- Age: mean 60 years (standard deviation 4.3)
- Gender: 63% male
- Ethnicity: not stated
- Smoking status: 42% ever smokers
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: not stated
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results: not stated
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: not stated
- Use of other therapy: not stated

### Outcomes
- Proportion of mortality

### Notes
- Idiopathic pulmonary fibrosis-related mortality
- Funding source: none

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**Zurkova 2016 [175]**
**Study Design**
- Type of Study: retrospective cohort
- Trial Design: national using the Czech Idiopathic Pulmonary Fibrosis Registry
- Country/ies: Czech Republic

**Study Duration**
1 year

**Participants**
- Number: 124
- Definition of diagnosis: unclear
- Age: not stated
- Gender: not stated
- Ethnicity: not stated
- Smoking status: not stated
- Use of home oxygen therapy: not stated
- Time since diagnosis: not stated
- Percentage of patients with surgical lung biopsy: not stated
- Percentage of patients with definite usual interstitial pneumonia patterns on HRCT: not stated
- Lung function results: not stated
- Resting oxyhaemoglobin saturation: not stated
- 6-minute walk distance: not stated
- Symptom assessment: not stated
- Use of systemic corticosteroid therapy: not stated
- Use of other therapy: not stated

**Outcomes**
- Change in forced vital capacity
- Change in diffusing capacity for carbon monoxide

**Notes**
Funding source: unclear
S6. Cohort studies: Documented treatments for IPF at baseline and/or during the study

| Study                  | Corticosteroids | Other Immunosuppressive agents | Colchicine | NAC | Investigational agents | Unclear | Other                          |
|------------------------|-----------------|--------------------------------|------------|-----|------------------------|---------|--------------------------------|
| Agusti 1993            | X               | X                              |            |     |                        |         | Inhaled bronchodilators        |
| Agusti 1994            | X               | X                              |            |     |                        |         |                                |
| Akagi 2009             | X               | X                              |            |     |                        |         |                                |
| Alhamad 2008           | X               | X                              | X          |     |                        |         | Oxygen therapy                 |
| Antoniou 2009          | X               | X                              |            |     |                        |         |                                |
| Araki 2003             | X               | X                              |            |     |                        |         | Oxygen therapy                 |
| Ashley 2016            | X               | X                              |            |     |                        |         |                                |
| Barlo 2009             | X               | X                              |            |     |                        |         |                                |
| Bennett 2015           | X               | X                              |            |     |                        |         | Doxycycline                     |
| Bhattacharyya 2009     | X               | X                              |            |     |                        |         |                                |
| Bjoraker 1998          | X               | X                              |            |     |                        |         |                                |
| Bjurstrom 2013         | X               | X                              |            |     |                        |         |                                |
| Boomars 1995           | X               | X                              |            |     |                        |         |                                |
| Bournazos 2011         | X               | X                              |            |     |                        |         |                                |
| Cai 2014               | X               | X                              | X          |     |                        |         | Oxygen therapy, interferon-gamma |
| Castria 2012           | X               | X                              |            |     |                        |         |                                |
| Cayon 2010             | X               | X                              |            |     |                        |         |                                |
| Civic 2012             | X               | X                              |            |     |                        |         |                                |
| Collins 2015           | X               | X                              |            |     |                        |         |                                |
| Cottin 2017            | X               | X                              |            |     |                        |         |                                |
| Couluris 2012          | X               | X                              |            |     |                        |         | Losartan, oxygen therapy       |
| Diaz 2012              | X               | X                              |            |     |                        |         |                                |
| Erbes 1997             | X               | X                              |            |     |                        |         |                                |
| Faverio 2015           | X               | X                              |            |     |                        |         |                                |
| Fiorucci 2008          | X               | X                              | X          |     |                        |         |                                |
| Fireman 1998           | X               | X                              |            |     |                        |         |                                |
| Fisher 2017a           | X               | X                              |            |     |                        |         |                                |
| Fisher 2017b           | X               | X                              |            |     |                        |         | Oxygen therapy                 |
| Fujimoto 2012          | X               | X                              |            |     |                        |         |                                |
| Gay 1998               | X               | X                              |            |     |                        |         |                                |
| Study                      | Corticosteroids | Other Immunosuppressive agents | Colchicine | NAC | Investigational agents | Unclear | Other                     |
|---------------------------|-----------------|--------------------------------|------------|-----|------------------------|---------|---------------------------|
| Gu 2014                   | X               |                                |            |     |                        |         |                           |
| Hallstrand 2005           | X               |                                |            |     |                        |         |                           |
| Hamada 2007               |                 |                                |            |     |                        |         |                           |
| Hanson 1995               | X               |                                |            |     |                        |         |                           |
| Harris 2010               |                 |                                |            |     |                        |         |                           |
| Hiwatari 1997             |                 |                                |            |     |                        |         |                           |
| Holland 2013              |                 |                                |            |     |                        |         |                           |
| Hopkins 2016              |                 |                                |            |     |                        |         |                           |
| Hosein 2016               |                 |                                |            |     |                        |         | Oxygen therapy            |
| Hubbard 1998              | X               |                                |            |     |                        |         |                           |
| Huynh 2015                |                 |                                |            |     |                        |         |                           |
| Inase 2003                | X               |                                |            |     |                        |         |                           |
| Iwasawa 2014              | X               |                                |            |     |                        |         |                           |
| Izumi 1992                | X               |                                |            |     |                        |         |                           |
| Jacob 2016                |                 |                                |            |     |                        |         |                           |
| Jaffar 2014               |                 |                                |            |     |                        |         |                           |
| Jeon 2006                 | X               |                                |            |     |                        |         | Interferon-gamma          |
| Jezek 1979                |                 |                                |            |     |                        |         |                           |
| Jezek 1980                | X               |                                |            |     |                        |         |                           |
| Jezkova 1981              |                 |                                |            |     |                        |         |                           |
| Jo 2017                   | X               |                                |            |     |                        |         |                           |
| Justet 2017               |                 |                                |            |     |                        |         | None at baseline          |
| Kanematsu 1994            |                 |                                |            |     |                        |         |                           |
| Khadadah 2003             | X               |                                |            |     |                        |         |                           |
| Kim 2012                  | X               |                                |            |     |                        |         |                           |
| Kim 2013                  |                 |                                |            |     |                        |         |                           |
| Kolb 1998                 | X               |                                |            |     |                        |         |                           |
| Kondoh 2005               | X               |                                |            |     |                        |         |                           |
| Kondoh 2010               |                 |                                |            |     |                        |         |                           |
| Korthagen 2011            | X               |                                |            |     |                        |         |                           |
| Kotecha 2016              | X               |                                |            |     |                        |         | Oxygen therapy            |
| Kreuter 2016              | X               |                                |            |     |                        |         |                           |
| Study                | Corticosteroids | Other Immunosuppressive agents | Colchicine | NAC | Investigational agents | Unclear | Other                        |
|---------------------|-----------------|--------------------------------|------------|-----|------------------------|---------|------------------------------|
| Kurashima 2010      | X               |                                 |            |     |                        |         | Inhaled bronchodilators     |
| Le Rouzic 2015      |                 |                                 |            |     |                        |         | X                           |
| Lederer 2006        |                 |                                 |            |     |                        |         | X                           |
| Lee 2011            | X               |                                 |            |     |                        |         | X                           |
| Lee 2012            | X               |                                 |            |     |                        |         | X                           |
| Lee 2015            | X               |                                 |            |     |                        |         | X                           |
| Li 2010             | X               |                                 |            |     |                        |         | X                           |
| Li 2012             |                 |                                 |            |     |                        |         | X                           |
| Li 2015             |                 |                                 |            |     |                        |         | X                           |
| Lindell 2015        |                 |                                 |            |     |                        |         | X                           |
| Liu 2017            |                 |                                 |            |     |                        |         | Oxygen therapy              |
| Lutherer 2011       |                 |                                 |            |     |                        |         | Interferon-gamma            |
| Mapel 1998          |                 |                                 |            |     |                        |         | X                           |
| Marulli 2010        |                 |                                 |            |     |                        |         | X                           |
| Mason 2007          |                 |                                 |            |     |                        |         | X                           |
| McKeown 2009        | X               |                                 |            |     |                        |         | X                           |
| Meier-Sydow 1979    | X               |                                 |            |     |                        |         | X                           |
| Meier-Sydow 1990    | X               |                                 |            |     |                        |         | X                           |
| Mermigkis 2015      | X               |                                 |            |     |                        |         | X                           |
| Mirrani 2012        |                 |                                 |            |     |                        |         | None at baseline             |
| Moeller 2009        | X               |                                 |            |     |                        |         | X                           |
| Mogulkoc 2001       | X               |                                 |            |     |                        |         | X                           |
| Moore 2013          |                 |                                 |            |     |                        |         | X                           |
| Morawiec 2011       |                 |                                 |            |     |                        |         | X                           |
| Moua 2016           | X               |                                 |            |     |                        |         | X                           |
| Mura 2012           |                 |                                 |            |     |                        |         | X                           |
| Nadrous 2004        | X               |                                 |            |     |                        |         | X                           |
| Nagai 1998          | X               |                                 |            |     |                        |         | X                           |
| Nambiar 2017        | X               |                                 |            |     |                        |         | Oxygen therapy              |
| Natsuzaka 2014      |                 |                                 |            |     |                        |         | X                           |
| Nicholson 2000      | X               |                                 |            |     |                        |         | X                           |
| Nicol 2015          |                 |                                 |            |     |                        |         | X                           |
| Nishiyama 2016      |                 |                                 |            |     |                        |         | Oxygen therapy              |
| Study                  | Corticosteroids | Other Immunosuppressive agents | Colchicine | NAC | Investigational agents | Unclear | Other                      |
|-----------------------|-----------------|---------------------------------|------------|-----|------------------------|---------|----------------------------|
| Nishiyama 2016b       |                 |                                 |            |     |                        | None at baseline |
| Oda 2014              |                 | X                               |            |     |                        | X       |                            |
| Oldham 2015           |                 | X                               | X          |     |                        | X       |                            |
| Prasse 2009           | X               | X                               |            |     |                        |         |                            |
| Raghu 2014            |                 | X                               |            |     |                        | X       | Interferon-gamma            |
| Raimundo 2016         |                 |                                 |            |     |                        | X       |                            |
| Rajasekaran 2006      | X               | X                               |            |     |                        |         |                            |
| Rangappa 2009         | X               | X                               |            |     |                        |         |                            |
| Reid 2015             |                 | X                               | X          |     |                        |         | Everolimus                  |
| Richards 2012         | X               | X                               | X          |     |                        |         | Interferon-gamma, tacrolimus |
| Rogers 2016           |                 |                                 |            |     |                        | X       |                            |
| Roig 2010             | X               | X                               |            |     |                        |         |                            |
| Roskell 2014          |                 |                                 |            |     |                        | X       |                            |
| Rubin 2015            |                 |                                 |            |     |                        | X       |                            |
| Rudd 2007             |                 | X                               |            |     |                        |         |                            |
| Russell 2016          |                 |                                 |            |     |                        | X       | Oxygen therapy              |
| Ryerson 2014          |                 |                                 |            |     |                        | X       |                            |
| Sakamoto 2010         | X               | X                               |            |     |                        |         |                            |
| Sato 2016             |                 |                                 |            |     |                        | X       |                            |
| Saydain 2002          |                 | X                               |            |     |                        |         |                            |
| Schmidt 2011          |                 |                                 |            |     |                        |         |                            |
| Schupp 2015           |                 |                                 |            |     |                        | X       |                            |
| Selman 1998           | X               | X                               |            |     |                        |         |                            |
| Selman 2007           | X               | X                               |            |     |                        |         |                            |
| Serrano-Mollan 2016   |                 |                                 |            |     |                        | X       | Prednisolone, azathioprine, NAC bosentan at baseline; none during study |
| Shafiq 2010           |                 |                                 |            |     |                        | X       |                            |
| Sherbini 2014         |                 | X                               | X          |     |                        |         | Oxygen therapy              |
| Shin 2008             | X               | X                               |            |     |                        | X       | Interferon-gamma            |
| Shinoda 2009          |                 |                                 |            |     |                        | X       |                            |
| Shitrit 2009          |                 | X                               |            |     |                        | X       | Oxygen therapy              |
| Shlobin 2009          |                 |                                 |            |     |                        | X       |                            |
| Study                  | Corticosteroids | Other Immunosuppressive agents | Colchicine | NAC | Investigational agents | Unclear | Other |
|-----------------------|-----------------|---------------------------------|------------|-----|------------------------|---------|-------|
| Siemienowicz 2015     |                 |                                 |            |     |                        | X       |       |
| Soares Pires 2013     | X               |                                 | X          | X   |                        |         |       |
| Soares 2015           |                 |                                 |            |     |                        |         |       |
| Sobiecka 2013         | X               |                                 |            | X   |                        |         |       |
| Song 2011             |                 |                                 |            |     |                        | X       |       |
| Song 2014             |                 |                                 |            |     |                        | X       |       |
| Stack 1972            |                 |                                 |            |     |                        | X       |       |
| Strand 2014           |                 |                                 |            |     |                        | X       |       |
| Strongman 2018        |                 |                                 |            |     |                        | X       |       |
| Su 2011               | X               |                                 |            |     |                        | X       |       |
| Takahashi 2000        |                 |                                 |            |     |                        | X       |       |
| ten Klooster 2015     |                 |                                 |            |     |                        | X       |       |
| Tokgoz Akyil 2016     |                 |                                 |            |     |                        | X       |       |
| Tolle 2014            |                 |                                 |            |     |                        | X       |       |
| Tomioka 2007          | X               |                                 |            |     |                        |         | Oxygen therapy |
| Tryfon 2009           |                 |                                 |            |     |                        | X       |       |
| Tzouvelekis 2013      | X               |                                 |            |     |                        | X       |       |
| Tzouvelekis 2016      |                 |                                 |            |     |                        |         | Oxygen therapy |
| Umeda 2015            |                 |                                 |            |     |                        |         | None at baseline |
| Vysehradsky 2002      | X               |                                 | X          |     |                        | X       |       |
| Wallaert 2011         |                 |                                 |            |     |                        | X       |       |
| Wilkie 2012           |                 |                                 |            |     |                        | X       |       |
| Wright 1981           |                 |                                 |            |     |                        | X       |       |
| Yokoyama 2006         |                 |                                 |            |     |                        | X       |       |
| Yu 2014               |                 |                                 |            |     |                        |         | Oxygen therapy |
| Zhang 2016            |                 |                                 |            |     |                        | X       |       |
| Zurkova 2016          | X               |                                 | X          |     |                        | X       |       |

Abbreviations: NAC, N-acetylcysteine
### S7. Randomised controlled trial: Concomitant treatment of IPF which were not allowed during the study

| Study          | Corticosteroids | Other Immunosuppressive agents | Antifibrotic drugs | NAC | Investigational agents | Additional notes                                                                                                                                 |
|----------------|-----------------|--------------------------------|--------------------|-----|------------------------|---------------------------------------------------------------------------------------------------------------------------------------------------|
| Daniels 2010   | X*              |                                |                    |     |                        | A 4-week washout period was required if participants were taking any prohibited concomitant treatment                                                |
| Demedts 2005   | ≥ 0.5 mg/kg/day of prednisone^ | ≥ 2 mg/kg/day of azathioprine^ | > 600mg/day for > 3 months in previous 3 years |     |                        | Treatment at study entry or in the past with drugs that interfere with the diagnosis, severity, therapy, or prognosis of IPF                      |
| King 2008      | Unstable dose or > 15 mg of prednisone or equivalent |                                |                   |     |                        |                                                                                                                                                    |
| King 2009      | 0.125 mg/kg /day or 0.25 mg/kg of prednisone every other day at randomisation |                                |                   |     | X^                     | Previous treatment with interferon gamma-1b was prohibited                                                                                         |
| King 2011      | > 20 mg per day prednisone or equivalent^ |                                |                   |     | X^                     |                                                                                                                                                    |
| King 2014      | X^              |                                |                    |     | X^                     | • Any cytotoxic, cytokine modulating, or receptor antagonist agents were prohibited  
• Medications that are specifically used for the treatment of IPF were prohibited*                                                                                        |
| Malouf 2011    | > 10mg/day of prednisolone |                                |                   |     |                        | Bosentan                                                                                                                                           |
| Martinez 2014  | As part of triple therapy | As part of triple therapy | As part of triple therapy |     |                        | • History of triple therapy for > 12 weeks’ duration in the past 4 years.  
• Triple therapy of ≤ 12 weeks duration in the past 4 years required a 30-day washout period before randomization.  
• Any therapy directed at pulmonary fibrosis (excepting triple therapy) required a 30-day washout period before randomized. |
| Study          | Corticosteroids | Other Immunosuppressive agents | Antifibrotic drugs | NAC | Investigational agents | Additional notes                                                                                                                                 |
|---------------|-----------------|--------------------------------|--------------------|-----|------------------------|----------------------------------------------------------------------------------------------------------------------------------------------------|
| Noble 2011    | X               | X                              | X                  | X   | X                      | Exceptions for short courses of azathioprine, cyclophosphamide, corticosteroids, or acetylcysteine for protocol-defined acute exacerbation of IPF, acute respiratory decompensation, or progression of disease. |
| Parker 2016   | > 15mg/day of prednisone or equivalent^ | X^                           | X^                 | X^  | X                      | Long-term use of phosphodiesterase-5 inhibitors for pulmonary hypertension was prohibited.                                                                 |
| Raghu 2013a   |                 | X^                            |                    |     |                         |                                                                                                                                                    |
| Raghu 2013b   |                 | X^                            | Calcineurin or mammalian target of rapamycin inhibitors^        | X^  |                         |                                                                                                                                                    |
| Raghu 2015    |                 | X                             |                    |     |                         | • Stable doses must have been established for all concomitant medications before and maintained throughout the treatment period. The initiation of new concomitant medications 6 for IPF was strongly discouraged through week 52. |
| Richeldi 2011 |                 | X                             |                    |     |                         | • Continuous oxygen therapy (> 15 hours/day) were ineligible. Concomitant anticoagulation medication was prohibited                                       |
| Richeldi 2014 | > 15 mg/day of prednisone or equivalent^ | X^                           | X^                 | X^  | X                      |                                                                                                                                                    |
| Taniguchi 2010 | > 10 mg/day of prednisone or equivalent | X                             |                    |     | X                      |                                                                                                                                                    |
S8. Risk of bias assessment

Table E5: Risk of bias assessment for included studies
(a) Cohort studies (n = 155; Mura et al.67 consisted of a retrospective and a prospective studies)

| Criteria                                      | High risk of bias | Low risk of bias | Unclear |
|-----------------------------------------------|-------------------|------------------|---------|
|                                               | Number of studies | %                | Number of studies | %          | Number of studies | %        |
| Clinical vs population sampling               | 150               | 97               | 4        | 2.5        | 1               | 0.5      |
| Prospective vs retrospective recruitment     | 99                | 64               | 55       | 35.5       | 1               | 0.5      |
| Selection criteria for participants described| 6                 | 4                | 135      | 87         | 14              | 9        |
| Baseline characteristics of participants described | 12             | 8                | 136      | 87         | 8               | 5        |
| Follow-up duration adequate                  | 0                 | 0                | 155      | 100        | 0               | 0        |
| Reason lost to follow-up described           | 16                | 10.5             | 16       | 10.5       | 123             | 79       |
| Timing of diagnosis at baseline              | 0                 | 0                | 154      | 99.5       | 1               | 0.5      |
| Outcome described a priori                   | 1                 | 0.5              | 142      | 91.5       | 12              | 8        |
| Adequate description of statistical analysis | 18                | 11.5             | 136      | 88         | 1               | 0.5      |

(b) RCTs (16 studies)

| Criteria                                      | High risk of bias | Low risk of bias | Unclear |
|-----------------------------------------------|-------------------|------------------|---------|
|                                               | Number of studies | %                | Number of studies | %          | Number of studies | %        |
| Clinical vs population sampling               | 16                | 100              | 0        | 0          | 0               | 0        |
| Selection criteria for participants described | 0                 | 0                | 16       | 100        | 0               | 0        |
| Baseline characteristics of participants described | 0              | 0                | 16       | 100        | 0               | 0        |
| Follow-up percentage adequate                | 1                 | 6                | 13       | 81         | 2               | 13       |
| Follow-up duration adequate                  | 0                 | 0                | 16       | 100        | 0               | 0        |
| Reason lost to follow-up described           | 1                 | 6                | 14       | 88         | 1               | 6        |
| Timing of diagnosis at baseline              | 0                 | 0                | 16       | 100        | 0               | 0        |
| Blinding described                            | 0                 | 0                | 13       | 81         | 3               | 19       |
| Outcome described a priori                   | 0                 | 0                | 16       | 100        | 0               | 0        |
| Use of intention-to-treat analysis           | 0                 | 0                | 13       | 81         | 3               | 19       |
| Adequate description of statistical analysis | 0                 | 0                | 16       | 100        | 0               | 0        |
Figure E1. Summary of risk of bias (as percentages across all included studies)
S9. Figures for results

Figure E2: Pooled mean changes in forced vital capacity (in litres) at 1 year to < 2 years

Figure E3: Pooled mean changes in diffusing capacity for carbon monoxide (in mL/mmHg/min) at 1 year to < 2 years
Figure E4: Pooled mean changes in 6MWD at 1 year to < 2 years
Figure E5: Subgroup analysis of pooled proportions of mortality at 1 year to < 2 years: Randomised controlled trials versus cohort studies

| Study                        | ES (95% CI) | % Weight |
|------------------------------|-------------|----------|
| Cohort study                 |             |          |
| Agadi 1993                   | 0.00 (0.00, 0.31) | 0.05     |
| Paonon 1995                  | 0.36 (0.16, 0.46) | 1.78     |
| Fienne 1998                  | 0.10 (0.03, 0.23) | 1.53     |
| Koub 1998                    | 0.33 (0.13, 0.35) | 1.4     |
| Mapel 1998                   | 0.22 (0.16, 0.28) | 1.93     |
| Kharah 2003                  | 0.07 (0.01, 0.24) | 1.36     |
| Kontoh 2003                  | 0.11 (0.02, 0.29) | 1.34     |
| Tomicka 2007                 | 0.27 (0.15, 0.43) | 1.55     |
| Akiaki 2009                  | 0.07 (0.02, 0.15) | 1.73     |
| Barlo 2009                   | 0.00 (0.00, 0.03) | 0.83     |
| Bhelacharanya 2009           | 0.29 (0.04, 0.71) | 0.69     |
| Shirl 2009                   | 0.06 (0.01, 0.16) | 1.60     |
| Shitom 2009                  | 0.20 (0.10, 0.22) | 2.05     |
| Caym 2013                    | 0.17 (0.02, 0.49) | 0.94     |
| March 2010                   | 0.21 (0.12, 0.34) | 1.64     |
| Lee 2011                     | 0.17 (0.10, 0.27) | 1.75     |
| Lutha 2011                   | 0.00 (0.00, 0.03) | 0.94     |
| Linhji 2011                  | 0.25 (0.19, 0.32) | 1.92     |
| Su 2011                      | 0.16 (0.10, 0.23) | 1.44     |
| Cic 2012                     | 0.09 (0.03, 0.17) | 1.54     |
| Diaz 2012                    | 0.00 (0.00, 0.03) | 0.84     |
| Li 2012                      | 0.47 (0.28, 0.66) | 1.39     |
| Mura 2012                    | 0.26 (0.16, 0.38) | 1.71     |
| Wikos 2012                   | 0.10 (0.05, 0.19) | 1.77     |
| Tzouvelie 2013               | 0.00 (0.00, 0.03) | 1.02     |
| Cai 2014                     | 0.39 (0.30, 0.46) | 1.93     |
| Gu 2014                      | 0.04 (0.00, 0.20) | 1.30     |
| Iwasawa 2014                 | 0.00 (0.00, 0.09) | 1.51     |
| Jaffar 2014                  | 0.29 (0.04, 0.71) | 0.69     |
| Shetani 2014                 | 0.30 (0.22, 0.39) | 1.86     |
| Song 2014                    | 0.40 (0.25, 0.56) | 1.54     |
| Yu 2014                      | 0.15 (0.07, 0.26) | 1.69     |
| Rennell 2015                 | 0.00 (0.00, 0.05) | 1.73     |
| Kim 2015                     | 0.10 (0.07, 0.15) | 1.96     |
| Lendell 2015                 | 0.08 (0.06, 0.12) | 1.99     |
| Ohtan 2015                   | 0.12 (0.08, 0.18) | 1.91     |
| Svirnenowicz 2015            | 0.35 (0.15, 0.59) | 1.20     |
| Ashley 2016                  | 0.00 (0.00, 0.05) | 1.66     |
| Knir 2016                    | 0.21 (0.16, 0.27) | 1.94     |
| Nalayama 2016                | 0.12 (0.02, 0.30) | 1.32     |
| Raimundo 2016                | 0.27 (0.20, 0.33) | 2.07     |
| Russell 2016                 | 0.26 (0.20, 0.42) | 1.60     |
| Serrano-Moller 2016          | 0.00 (0.00, 0.02) | 1.09     |
| Tzouvelie 2016               | 0.11 (0.06, 0.17) | 1.79     |
| Fisher 2017                  | 0.10 (0.07, 0.14) | 1.97     |
| Strongman 2018               | 0.28 (0.27, 0.30) | 2.06     |
| Overall (I² = 93.40%, p = 0.00) | 0.14 (0.12, 0.17) | 71.90     |

| RCT                          |             |          |
| Darwood 2005                 | 0.11 (0.05, 0.20) | 1.73     |
| King 2008                    | 0.10 (0.04, 0.18) | 1.75     |
| King 2009                    | 0.14 (0.10, 0.19) | 1.96     |
| Daniel 2010                  | 0.17 (0.08, 0.29) | 1.65     |
| Taniguchi 2010               | 0.01 (0.00, 0.05) | 0.81     |
| King 2011                    | 0.03 (0.01, 0.06) | 1.93     |
| Malan 2011                   | 0.13 (0.05, 0.27) | 1.56     |
| Rulsee 2011                  | 0.08 (0.06, 0.12) | 1.58     |
| Ruchal 2011                  | 0.11 (0.05, 0.19) | 1.76     |
| Ruphi 2013a                  | 0.04 (0.01, 0.08) | 1.90     |
| Ruphi 2013b                  | 0.02 (0.00, 0.09) | 1.65     |
| King 2014                    | 0.07 (0.04, 0.11) | 1.96     |
| Martinez 2014                | 0.02 (0.00, 0.07) | 1.86     |
| Ruchal 2014                  | 0.08 (0.05, 0.11) | 2.00     |
| Ruphi 2015                   | 0.11 (0.02, 0.29) | 1.96     |
| Overall (I² = 75.01%, p = 0.00) | 0.07 (0.05, 0.09) | 28.50     |

Proportion of mortality

Overall (I² = 95.12%, p = 0.00): 0.12 (0.09, 0.14) 100.00
Figure E6: Sensitivity analyses of pooled proportions of mortality at different time frames

a) At one year to less than two years

b) Between two and five years
c) Five years or greater

| Study       | ES (95% CI)         | Weight |
|-------------|---------------------|--------|
| Jeon 2006   | 0.59 (0.48, 0.69)   | 11.04  |
| Hamada 2007 | 0.33 (0.21, 0.46)   | 10.77  |
| Shin 2008   | 0.54 (0.43, 0.66)   | 10.97  |
| Akagi 2009  | 0.76 (0.65, 0.85)   | 10.95  |
| Barfo 2009  | 0.96 (0.90, 0.99)   | 11.18  |
| Su 2011     | 0.48 (0.38, 0.57)   | 11.22  |
| Cai 2014    | 0.61 (0.54, 0.68)   | 11.42  |
| Oldham 2015 | 0.44 (0.37, 0.52)   | 11.38  |
| Zhang 2016  | 0.34 (0.25, 0.45)   | 11.06  |
| Overall     | 0.56 (0.43, 0.71)   | 100.00 |
S10. Additional results

i. Progression-free survival
It was not possible to perform pooled analysis of progression-free survival due to variations in the definition of disease progression for each study. Two studies defined disease progression as death or a decline in lung function.\(^8,16^7\) Kotecha et al reported a median progression-free survival of 13 months in 27 patients with a median follow-up duration of 33 months,\(^8\) while Umeda et al reported a median progression-free survival of 27.9 months in 23 patients after five years of follow-up.\(^16^7\) After two years of follow-up, Prasse et al reported median times to decline in lung function of 6 to 15 months in 72 patients.\(^12^1\) Richards et al reported median times to decline in lung function of 1.01 to 1.05 years in 241 patients with a median follow-up duration of 1.4 to 1.8 years.\(^12^7\)

ii. Respiratory-related mortality
Data were available for pooling from seven studies with a total of 1,094 participants.\(^15,17^5,17^9,1^7^6,1^7^8,1^8^3,1^8^4\) The pooled proportion of respiratory-related mortality at one year to less than two years was 0.06 (95% CI: 0.03 to 0.09) (Figure S1). There was moderately significant heterogeneity among pooled studies. Over three years of follow-up in the study by Roig et al with 46 participants, 18 (39%) died from respiratory-related causes.\(^12^9\)

![Figure E7. Pooled proportion of respiratory-related mortality at 1 year to < 2 years](image)

IPF-related mortality was reported as an outcome measure in three studies.\(^1^4^3,1^7^4,1^8^3\) King et al provided pooled data of two RCTs for IPF-related mortality of 3.5% at 12 months,\(^1^4^3,1^8^3\) with a total of 624 participants. Zhang et al reported IPF-related mortality of 23.3% at five years, in a study of 90 participants.\(^1^7^4\)
iii. **Proportion of patients with an absolute decline in FVC of ≥ 10% predicted**

At one year, the pooled proportion of patients with an absolute decline in FVC at 10% predicted or more was 0.35 (95% CI: 0.28 to 0.43; 8 studies, 1076 participants) (Figure S4). There was moderately significant heterogeneity among pooled studies ($I^2 = 81\%$, $p < 0.0001$).

| Study          | ES (95% CI) | Weight |
|----------------|------------|--------|
| Hanson 1995    | 0.24 (0.14, 0.37) | 11.68  |
| Kondoh 2005    | 0.33 (0.17, 0.54) | 8.56   |
| Taniguchi 2010 | 0.52 (0.42, 0.62) | 13.61  |
| King 2014      | 0.32 (0.26, 0.38) | 15.66  |
| Oda 2014       | 0.19 (0.12, 0.29) | 13.43  |
| Richeldi 2014  | 0.39 (0.35, 0.44) | 16.17  |
| Parker 2016    | 0.49 (0.36, 0.63) | 11.91  |
| Nambiar 2017   | 0.38 (0.21, 0.56) | 9.28   |
| Overall ($I^2 = 80.72\%$, $p = 0.00$) | 0.35 (0.28, 0.43) | 100.00 |

Figure E8. Pooled proportion of patients with an absolute decline in FVC of 10% predicted or more at 1 year to < 2 years

iv. **Proportion of patients with an absolute decline in DLCO of ≥ 15% predicted**

Although no study reported the proportion of patients with an absolute decline of 15% predicted or more in DLCO, there were three studies which classified the decline in DLCO at one year using different parameters. Demedts et al found 51% of 63 participants experienced a decline in DLCO of > 15% predicted or 1 mmol/min/kPa. Hanson et al reported 10 (23%) participants had a reduction in DLCO of at least 20%. In a study of 144 participants by Schmidt et al, there were 57 participants (39.6%) with a relative decline in DLCO of at least 15%.

v. **Change in dyspnoea**

King et al similarly found a worsening of symptoms over 12 months with a mean change of -1.7 (standard deviation = 3.6) in the Transition Dyspnoea Index.

vi. **Change in health-related quality of life using the EQ-5D**

Due to differences in reported methods, a pooled analysis could not be performed for the two studies which measured changes in health-related quality of life using the EQ-5D. In a study of 131 patients, Martinez et al reported a decline in health-related quality of life with a mean change of -3.3 for the EQ-5D visual analogue score at 60 weeks. Parker et al found a mean change of -0.815 for the EQ-5D utility index in 57 patients at 72 weeks.