Supplementary file 1: Treatment SLR EGPA, SLR Diagnostic & Follow-up, SLR General Management

Online supplementary material

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Supplementary file 1: Treatment SLR EGPA, SLR Diagnostic & Follow-up, SLR General Management

1. Research questions

14 research questions were developed. The PICO questions addressing treatment of EGPA (#11 to #13) diagnostic procedures and follow-up testing (#1, #2, #14) and supplementary search for areas of general management (without PICO questions) are addressed in this article, research questions addressing treatment of GPA and MPA are addressed in the complementary SLR article.

1.1. Research questions in PICO format addressing treatment of EGPA.

<table>
<thead>
<tr>
<th>PICO #</th>
<th>PICO question</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td><strong>Remission induction (immunosuppressive drug treatment)</strong></td>
</tr>
</tbody>
</table>
| PICO #11| In patients with new-onset and/or relapsing active **severe** EGPA (P), what is the impact of cyclophosphamide or other immunosuppressive drugs (I) vs. comparator immunosuppressive drugs such as rituximab (C1), or other drugs (C2,…) on disease-related outcomes (O1) and treatment-related adverse events (O2)?  
- Outcomes to consider: disease activity, disease damage, relapse, death, recovery of renal function, control of asthma symptoms, control of rhino-sinusitis symptoms, health-related quality of life and other patient-related outcomes, infection, malignancy including bladder cancer, serious adverse events, hypogammaglobulinemia, toxicity leading to discontinuation  
- Interventions and comparators to consider: Cyclophosphamide (oral and intravenous), Rituximab, mycophenolate mofetil, IVIG, azathioprine, methotrexate, leflunomide, mepolizumab, benralizumab, omalizumab  |
| PICO #12| In patients with new-onset and/or relapsing active **non-severe** EGPA (P), what is the impact of mepolizumab or immunosuppressive drugs (I) vs. comparator immunosuppressive drugs such as rituximab or GC monotherapy (C1), or other drugs (C2,…) on disease-related outcomes (O1) and treatment-related adverse events (O2)?  
- Outcomes to consider: disease activity, disease damage, relapse, death, recovery of renal function, control of asthma symptoms, control of rhino-sinusitis symptoms, health-related quality of life and other patient-related outcomes, infection, malignancy including bladder cancer, serious adverse events, hypogammaglobulinemia, toxicity leading to discontinuation  
- Interventions and comparators to consider: Cyclophosphamide (oral and intravenous), Rituximab, mycophenolate mofetil, IVIG, azathioprine, methotrexate, leflunomide, mepolizumab, benralizumab, omalizumab.                                    |
|         | **Remission maintenance (immunosuppressive drug treatment)**                                                                                                                                                                    |
| PICO #13| In patients with GPA/MPA in remission after induction therapy (P), what is the impact of using azathioprine (I) vs. comparator immunosuppressive drugs (including MTX, rituximab and MMF)) (C ) on disease-related outcomes and treatment-related adverse events (O)?  
- Outcomes to consider: relapse (minor and major), time to first relapse, death, disease damage, renal function, infection, health-related quality of life and other patient-related outcomes, serious adverse events, toxicity leading to discontinuation, glucocorticoid use (cumulative dose)  
- Interventions and comparators to consider: Cyclophosphamide (oral and intravenous), Rituximab, mycophenolate mofetil, IVIG, azathioprine, methotrexate, leflunomide, mepolizumab, benralizumab, omalizumab. |
1.2. Research questions in PICO format addressing diagnostic procedures and follow-up testing

<table>
<thead>
<tr>
<th>PICO #</th>
<th>PICO question</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Diagnostic procedures</strong></td>
<td></td>
</tr>
<tr>
<td>PICO # 1</td>
<td>In patients with GPA/MPA (P1) or EGPA (P2), what is the impact of a positive tissue biopsies (I) vs obtaining no biopsy/negative biopsy (C) to confirm a clinical diagnosis of AAV (O)?</td>
</tr>
<tr>
<td>PICO # 2</td>
<td>In patients with GPA/MPA (P1) or EGPA (P2), what is the impact of a positive ANCA vs a negative ANCA (C) to confirm a clinical diagnosis of AAV (O)?</td>
</tr>
<tr>
<td><strong>Follow-up testing</strong></td>
<td></td>
</tr>
<tr>
<td>PICO # 14</td>
<td>In patients with AAV (P), what is the impact of measurement of which clinical parameters, tests and biomarkers (I) vs. not measuring these (C) on disease-related outcomes and treatment-related adverse events (O)?</td>
</tr>
<tr>
<td>- Outcomes to consider: relapse (minor and major), time to first relapse, death, disease damage, renal function, infection, health-related quality of life and other patient-related outcomes, serious adverse events, toxicity leading to discontinuation</td>
<td></td>
</tr>
</tbody>
</table>
Supplementary file 1: Treatment SLR EGPA, SLR Diagnostic & Follow-up, SLR General Management

2. Search strategies

2.1. Search strategy treatment PubMed

SeekBar("Anti-Neutrophil Cytoplasmic Antibody-Associated Vasculitis"[Mesh]) OR ("ANCA associated vasculitis"[tiab]) OR ("churg-strauss" [tiab]) OR ("wegener" [tiab]) OR ("granulomatosis"[tiab]) OR ("microscopic"[tiab]) AND ("polyangitis"[tiab]) OR ("ANCA"[tiab]) OR ("Antibodies, Antineutrophil Cytoplasmic"[Mesh]) OR ("antineutrophil cytoplasmic antibody"[tiab]) AND ("vasculitis"[tiab]) OR ("glomerulonephritis"[tiab]) OR ("renal"[tiab]))

AND

(("Drug Therapy"[Mesh]) OR ("therapy"[tiab]) OR ("treatment" [tiab]) OR ("Glucocorticoids" [Pharmacological Action]) OR ("Glucocorticoid" [tiab]) OR ("steroid" [tiab]) OR ("corticosteroid" [tiab]) OR ("prednisone"[tiab]) OR ("prednisolone"[tiab]) OR ("methylprednisolone"[tiab]) OR ("dexamethasone"[tiab]) OR ("Cyclophosphamide"[Mesh]) OR ("Cyclophosphamide" [tiab]) OR ("rituximab" [tiab]) OR ("methotrexate" [tiab]) OR ("azathioprine" [tiab]) OR ("immunosuppress*" [tiab]) OR ("leflunomide" [tiab]) OR ("cyclosporine" [tiab]) OR ("mycophenol*" [tiab]) OR ("Trimethoprim" [tiab]) OR ("Sulfamethoxazole" [tiab]) OR ("Cotrim" [tiab]) OR ("Immunoglobulins, Intravenous"[Mesh]) OR ("immunoglobulin*"[tiab]) OR ("biologic*" [tiab]) AND ("agent" [tiab]) OR ("therapy"[tiab])) OR ("plasmapheresis"[Mesh]) OR ("plasmapheresis" [tiab]) OR ("plasma exchange" [tiab]) OR ("trial"[tiab]) OR ("random*"[tiab]) OR ("Randomized Controlled Trial" [Publication Type]) OR ("Controlled Clinical Trial" [Publication Type]) AND ("2015/02/01"[Date - Publication] : "3000"[Date - Publication]) OR ("mepolizumab" [tiab]) OR ("reslizumab" [tiab]) OR ("benralizumab" [tiab]) OR ("interleukin 5"[tiab]) OR ("omalizumab"[tiab]) OR ("avacopan" [tiab]) OR ("CCX168"[tiab]) OR ("belimumab" [tiab]) OR ("abatacept" [tiab]) OR ("Janus Kinase Inhibitors"[Mesh]) OR ("Janus kinase inhibitor*" [tiab]) OR ("tofacitinib" [tiab]) OR ("baricitinib" [tiab]) OR ("upadacitinib" [tiab]) OR ("filgotinib" [tiab]))

NOT

("Case reports" [Publication Type])
2.2. Search strategy treatment EMBASE

('ANCA associated vasculitis'/exp OR 'ANCA associated vasculiti*':ab,ti OR 'churg-strauss':ab,ti OR "wegener":ab,ti OR ((granulomatosis:ab,ti OR microscopic:ab,ti) AND polyangiitis:ab,ti) OR ((ANCA:ab,ti OR 'neutrophil cytoplasmic antibody'/exp OR 'antineutrophil cytoplasmic antibod*':ab,ti) AND (vasculitis:ab,ti OR glomerulonephritis:ab,ti OR renal:ab,ti)))

AND

(((('drug therapy'/exp OR therapy:ab,ti OR treatment:ab,ti OR 'glucocorticoid'/exp OR glucocorticoid:ab,ti OR steroid:ab,ti OR corticosteroid:ab,ti OR prednisone:ab,ti OR prednisolone:ab,ti OR methylprednisolone:ab,ti OR dexamethasone:ab,ti OR 'cyclophosphamide'/exp OR cyclophosphamide:ab,ti OR rituximab:ab,ti OR methotrexate:ab,ti OR azathioprine:ab,ti OR immunosuppress*:ab,ti OR leflunomide:ab,ti OR cyclosporine:ab,ti OR Mycophenol*:ab,ti OR trimethoprim:ab,ti OR Sulfamethoxazole:ab,ti OR Cotrim:ab,ti OR 'immunoglobulin'/exp OR immunoglobulin*:ab,ti OR (biologic*:ab,ti OR AND (agent:ab,ti OR therapy:ab,ti)) OR 'plasmapheresis'/exp OR plasmapheresis:ab,ti OR 'plasma exchange':ab,ti OR trial:ab,ti OR random*:ab,ti OR 'Controlled Clinical Trial'/exp) AND [01-02-2015]/sd) OR (mepolizumab:ab,ti OR reslizumab:ab,ti OR benralizumab:ab,ti OR 'interleukin 5':ab,ti OR omalizumab:ab,ti OR avacopan:ab,ti OR CCX168:ab,ti OR belimumab:ab,ti OR abatacept:ab,ti OR 'janus kinase inhibitor'/exp OR 'janus kinase inhibitor*':ab,ti OR tofacitinib:ab,ti OR baricitinib:ab,ti OR upadacitinib:ab,ti OR filgotinib:ab,ti))

AND

([article]/lim OR [article in press]/lim OR [review]/lim)

NOT

'case report'/exp
2.3. Search strategy treatment Cochrane Library

ID   Search
#1   MeSH descriptor: [Anti-Neutrophil Cytoplasmic Antibody-Associated Vasculitis] explode all trees
#2   (ANCA associated vasculiti* OR churg-strauss OR wegener):ti,ab,kw (Word variations have been searched)
#3   (granulomatosis OR microscopic):ti,ab,kw (Word variations have been searched)
#4   (polyangiitis):ti,ab,kw (Word variations have been searched)
#5   #3 AND #4
#6   MeSH descriptor: [Antibodies, Antineutrophil Cytoplasmic] explode all trees
#7   (antineutrophil cytoplasmic antibod* OR ANCA):ti,ab,kw (Word variations have been searched)
#8   #6 OR #7
#9   (vasculitis OR glomerulonephritis OR renal):ti,ab,kw (Word variations have been searched)
#10  #8 AND #9
#11  #1 OR #2 OR #5 OR #10
#12  MeSH descriptor: [Drug Therapy] explode all trees
#13  MeSH descriptor: [Glucocorticoids] explode all trees
#14  MeSH descriptor: [Cyclophosphamide] explode all trees
#15  MeSH descriptor: [Immunoglobulins, Intravenous] explode all trees
#16  MeSH descriptor: [Plasmapheresis] explode all trees
#17  ("randomized controlled trial"):pt
#18  ("controlled clinical trial"):pt
#19  (biologic):ti,ab,kw (Word variations have been searched)
#20  (agent OR therapy):ti,ab,kw (Word variations have been searched)
#21  #19 AND #20
#22  (therapy OR treatment OR Glucocorticoid OR steroid OR corticosteroid OR prednisone OR prednisolone OR methylprednisolone OR dexamethasone OR cyclophosphamide OR rituximab OR methotrexate OR azathioprine OR immunosuppress*: OR leflunomide OR cyclosporine OR mycophenol*: OR Trimethoprim OR Sulfamethoxazole OR Cotrim OR immunoglobulin*: OR plasmapheresis OR plasma exchange OR trial OR random*:ti,ab,kw (Word variations have been searched)
#23  #12 OR #13 OR #14 OR #15 OR #16 OR #17 OR #18 OR #21 OR #22 with Cochrane Library publication date Between Feb 2015 and Feb 2022
#24  MeSH descriptor: [Janus Kinase Inhibitors] explode all trees
#25  (mepolizumab OR reslizumab OR benralizumab OR interleukin 5 OR omalizumab OR avacopan OR CCX168 OR belimumab OR abatacept OR Janus kinase inhibitor*: OR tofacitinib OR baricitinib OR upadacitinib OR filgotinib):ti,ab,kw (Word variations have been searched)
#26  #24 OR #25
#27  #23 OR #26
#28  #11 AND #27

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2.4. Search strategy Diagnostic procedures and follow-up testing PubMed

(("Anti-Neutrophil Cytoplasmic Antibody-Associated Vasculitis"[Mesh]) OR ("ANCA associated vasculitis"[tiab]) OR ("churg-strauss" [tiab]) OR ("wegener" [tiab]) OR ("granulomatosis"[tiab]) OR ("microscopic"[tiab]) AND ("polyanagitis"[tiab])) OR ("Antibodies, Antineutrophil Cytoplasmic"[Mesh]) OR ("antineutrophil cytoplasmic antibody"[tiab]) AND ("vasculitis"[tiab]) OR ("glomerulonephritis"[tiab]) OR ("renal"[tiab]))

AND

((("Antibodies, Antineutrophil Cytoplasmic"[Mesh]) OR ("antineutrophil cytoplasmic antibody"[tiab]) OR ("myeloperoxidase" [tiab]) OR ("proteinase 3" [tiab]) OR ("MPO" [tiab]) OR ("PR3" [tiab]) OR ("Autoantibodies"[Mesh]) OR ("Autoantibody"[tiab])) AND ("Imunoenzyme Techniques"[Mesh]) OR ("Immunosorbent Techniques"[Mesh]) OR ("Immunofluorescent Antigen Technique, Indirect"[Mesh]) OR ("Assay"[tiab]) OR ("ELISA"[tiab]) OR ("IFT"[tiab]) OR ("measur*"[tiab]) OR ("test*"[tiab]) OR ("immunosorbent"[tiab]) OR ("immunoenzyme"[tiab]) OR ("immunofluorescent"[tiab]) OR ("positive"[tiab]) OR ("negative"[tiab])) OR ("Pathology"[MeSH]) OR ("Pathology"[tiab]) OR ("Histology"[MeSH]) OR ("Histology"[tiab]) OR ("Biops*"[tiab]) OR ("Surrogate"[tiab]) OR ("Diagnostic Imaging"[MeSH]) OR ("Diagnostic Imaging"[tiab]) OR ("Diagnosis"[MeSH]) OR ("Diagnosis"[tiab]) OR ("Radiography, Thoracic"[Mesh]) OR ("x-ray"[tiab]) OR ("Tomography, X-Ray Computed"[MeSH]) OR ("Computed Tomography"[tiab]) OR ("CT"[tiab]) OR ("Ultrasonography"[MeSH]) OR ("Ultrasonography"[tiab]) OR ("Ultrasonograph"[tiab]) OR ("Magnetic Resonance Imaging"[MeSH]) OR ("Magnetic Resonance Imaging"[tiab]) OR ("MRI"[tiab]) OR ("PET"[tiab]) OR ("biomarkers"[MeSH]) OR ("genotype"[tiab]) OR ("Blood Cell Count"[Mesh]) OR ("b cell*"[tiab]) OR ("b lymphocyte*" [tiab]) OR ("eosinophil*"[tiab]) OR ("cell"[tiab]) AND ("count"[tiab]) OR ("Urinalysis"[Mesh]) OR ("Urinalysis"[tiab]) OR ("genotype"[Mesh]) OR ("Genotype"[tiab]) OR ("Genetic"[tiab]) OR ("Phenotype"[Mesh]) OR ("Phenotype"[tiab]) OR ("Serotype"[tiab]) OR ("Symptom Assessment"[MeSH]) OR ("Symptom"[tiab]) OR ("Physical Examination"[MeSH]) OR ("Physical Examination"[tiab]) OR ("Birmingham Vasculitis Activity Score"[tiab]) OR ("BVAS"[tiab]) OR ("Five factor score"[tiab]) OR ("Vasculitis Damage Index"[tiab]) OR ("AAV-PRO"[tiab]) OR ("Classification"[MeSH]) OR ("Classification"[tiab]))

AND

("2015/02/01"[Date - Publication] : "3000"[Date - Publication])

NOT

("Case reports" [Publication Type])
Supplementary file 1: Treatment SLR EGPA, SLR Diagnostic & Follow-up, SLR General Management

2.5. Search strategy Diagnostic procedures and follow-up testing EMBASE

('ANCA associated vasculitis'/exp OR 'ANCA associated vasculiti*':ab,ti OR 'churg-strauss':ab,ti OR wegener:ab,ti OR ((granulomatosis:ab,ti OR microscopic:ab,ti) AND polyangiitis:ab,ti) OR ((ANCA:ab,ti OR 'neutrophil cytoplasmic antibody'/exp OR 'antineutrophil cytoplasmic antibody*':ab,ti) AND (vasculitis:ab,ti OR glomerulonephritis:ab,ti OR renal:ab,ti))

AND

[['neutrophil cytoplasmic antibody'/exp OR 'antineutrophil cytoplasmic antibod*':ab,ti OR ANCA:ab,ti OR myeloperoxidase:ab,ti OR 'proteinase 3':ab,ti OR MPO:ab,ti OR PR3:ab,ti OR 'proteinase 3 neutrophil cytoplasmic antibody'/exp OR 'myeloperoxidase antibody'/exp OR 'autoantibody'/exp OR Autoantibod*:ab,ti) AND ('enzyme immunoassay'/exp OR 'enzyme linked immunosorbent assay'/exp OR 'indirect fluorescent antibody technique'/exp OR Assay:ab,ti OR ELISA:ab,ti OR IFT:ab,ti OR measur*:ab,ti OR test*:ab,ti OR immunosorbent:ab,ti OR immunoenzyme:ab,ti OR immunofluorescent:ab,ti OR positive:ab,ti OR negative:ab,ti)) OR 'pathology'/exp OR 'histology'/exp OR Pathology:ab,ti OR Histology:ab,ti OR 'biopsy'/exp OR Biops*:ab,ti OR Surrogate:ab,ti OR 'diagnostic imaging'/exp OR 'Diagnostic Imaging':ab,ti OR 'diagnostic procedure'/exp OR Diagnosis:ab,ti OR 'thorax radiography'/exp OR 'x-ray':ab,ti OR 'x-ray computed tomography'/exp OR 'Computed Tomography':ab,ti OR CT:ab,ti OR 'echography'/exp OR Ultrasonography:ab,ti OR Ultrasound:ab,ti OR 'nuclear magnetic resonance imaging'/exp OR 'Magnetic Resonance Imaging':ab,ti OR MRI:ab,ti OR 'positron emission tomography'/exp OR PET:ab,ti OR 'biological markers'/exp OR biomarker*:ab,ti OR 'blood cell count'/exp OR 'b cell*':ab,ti OR 'b lymphocyte*':ab,ti OR ((eosinophil*:ab,ti OR cell:ab,ti) AND count:ab,ti) OR 'urine test strip'/exp OR urinalysis:ab,ti OR 'genotype'/exp OR genotype:ab,ti OR genetic:ab,ti OR 'phenotype'/exp OR Phenotype:ab,ti OR Serotype:ab,ti OR 'Symptom Assessment'/exp OR Symptom:ab,ti OR 'physical examinations and diagnoses'/exp OR 'Physical Examination':ab,ti OR 'Birmingham Vasculitis Activity Score':ab,ti OR BVAS:ab,ti OR 'Five factor score':ab,ti OR 'Vasculitis Damage Index':ab,ti OR 'AAV-PRO':ab,ti OR 'vasculitis damage index'/exp OR 'birmingham vasculitis activity score'/exp OR 'disease classification'/exp OR classification:ab,ti)

AND

[01-02-2015]/sd

AND

[(article)/lim OR [article in press]/lim OR [review]/lim]

NOT

'case report'/exp
2.6. Search strategy diagnostic procedures and follow-up testing Cochrane Library

ID #1 MeSH descriptor: [Anti-Neutrophil Cytoplasmic Antibody-Associated Vasculitis] explode all trees
#2 (ANCA associated vasculiti* OR churg-strauss OR wegener):ti,ab,kw (Word variations have been searched)
#3 (granulomatosis OR microscopic):ti,ab,kw (Word variations have been searched)
#4 (polyangiitis):ti,ab,kw (Word variations have been searched)
#5 #3 AND #4
#6 MeSH descriptor: [Antibodies, Antineutrophil Cytoplasmic] explode all trees
#7 (antineutrophil cytoplasmic antibod* OR ANCA):ti,ab,kw (Word variations have been searched)
#8 #6 OR #7
#9 (vasculitis OR glomerulonephritis OR renal):ti,ab,kw (Word variations have been searched)
#10 #8 AND #9
#11 #1 OR #2 OR #5 OR #10
#12 MeSH descriptor: [Antibodies, Antineutrophil Cytoplasmic] explode all trees
#13 MeSH descriptor: [Autoantibodies] explode all trees
#14 (antineutrophil cytoplasmic antibod* OR ANCA OR myeloperoxidase OR proteinase 3 OR MPO OR PR3 OR Autoantibod*):ti,ab,kw (Word variations have been searched)
#15 #12 OR #13 OR #14
#16 MeSH descriptor: [Immunoenzyme Techniques] explode all trees
#17 MeSH descriptor: [Immunosorbent Techniques] explode all trees
#18 MeSH descriptor: [Fluorescent Antibody Technique, Indirect] explode all trees
#19 (Assay OR ELISA OR IFT OR measur* OR test* OR immunosorbent OR immunoenzyme OR immunofluorescent OR positive OR negative):ti,ab,kw (Word variations have been searched)
#20 #15 OR #17 OR #18 OR #19
#21 #15 AND #20
#22 MeSH descriptor: [Pathology] explode all trees
#23 MeSH descriptor: [Histology] explode all trees
#24 MeSH descriptor: [Diagnostic Imaging] explode all trees
#25 MeSH descriptor: [Diagnosis] explode all trees
#26 MeSH descriptor: [Radiography, Thoracic] explode all trees
#27 MeSH descriptor: [Tomography, X-Ray Computed] explode all trees
#28 MeSH descriptor: [Ultrasonography] explode all trees
#29 MeSH descriptor: [Magnetic Resonance Imaging] explode all trees
#30 MeSH descriptor: [Positron-Emission Tomography] explode all trees
#31 MeSH descriptor: [Biomarkers] explode all trees
#32 MeSH descriptor: [Blood Cell Count] explode all trees
#33 MeSH descriptor: [Urinalysis] explode all trees
#34 MeSH descriptor: [Genotype] explode all trees
#35 MeSH descriptor: [Phenotype] explode all trees
#36 MeSH descriptor: [Symptom Assessment] explode all trees
#37 MeSH descriptor: [Physical Examination] explode all trees
#38 MeSH descriptor: [Classification] explode all trees
#39 (eosinophil* OR cell):ti,ab,kw (Word variations have been searched)
#40 (count):ti,ab,kw (Word variations have been searched)
#41 #39 AND #40
#42 (Pathology OR Histology OR Biops* OR Surrogate OR Diagnostic Imaging OR Diagnosis OR x-ray OR Computed Tomography OR CT OR Ultrasonography OR Ultrasound OR Magnetic Resonance Imaging OR MRI OR PET OR biomarker* OR b cell* OR b lymphocyte* OR Urinalysis OR Genotype OR Genetic OR Phenotype OR Serotype OR Symptom OR Physical Examination OR Birmingham Vasculitis Activity Score OR BVAS OR Five factor score OR Vasculitis Damage Index OR AAV-PRO OR classification):ti,ab,kw (Word variations have been searched)
#43 #21 OR #22 OR #24 OR #25 OR #26 OR #27 OR #28 OR #29 OR #30 OR #31 OR #32 OR #33 OR #34 OR #35 OR #36 OR #37 OR #38 OR #41 OR #42
#44 #11 AND #43 with Cochrane Library publication date Between Feb 2015 and Feb 2022
2.7. Search strategy General Management PubMed

(("Anti-Neutrophil Cytoplasmic Antibody-Associated Vasculitis"[Mesh]) OR ("ANCA associated vasculiti*" [tiab]) OR ("churg-strauss" [tiab]) OR ("wegener" [tiab]) OR ("granulomatosis"[tiab]) OR ("microscopic"[tiab]) AND ("polyangitis"[tiab]) OR ("[ANCA]"[tiab]) OR ("Antibodies, Antineutrophil Cytoplasmic"[Mesh]) OR ("antineutrophil cytoplasmic antibod*"[tiab]) AND ("vasculitis"[tiab]) OR ("glomerulonephritis"[tiab]) OR ("renal"[tiab])))

AND

(("Aged"[Mesh]) OR ("Aged"[tiab]) OR ("Child"[tiab]) OR ("Child"[Mesh]) OR ("Adolescent"[Mesh]) OR ("Adolescent"[tiab]) OR ("pregnancy"[tiab]) OR ("Contraception"[Mesh]) OR ("Contraception"[tiab]) OR ("Kidney Transplantation"[Mesh]) OR ("Kidney Transplantation"[tiab]) OR ("Antihypertensive Agents"[Mesh]) OR ("Antihypertensive"[tiab]) OR ("Rehabilitation"[Mesh]) OR ("Rehabilitation"[tiab]) OR ("patient care" [Mesh]) OR ("patient care" [tiab]) OR ("shared decision making" [tiab]) OR ("Shared Medical Appointments"[Mesh]) OR ("Shared Medical Appointment*"[tiab]) OR ("Patient Education as Topic"[Mesh]) OR ("Patient Education"[tiab]) OR ("Risk Reduction Behavior"[Mesh]) OR ("Risk Reduction"[tiab]) OR ("Patient Reported Outcome Measures"[Mesh]) OR ("Patient Reported Outcome" [tiab]) OR ("Fatigue"[tiab]) OR ("Smoking"[tiab]) OR ("Vaccination"[tiab]) OR ("SARS-CoV-2" [Mesh]) OR ("SARS-CoV-2" [tiab]) OR ("COVID-19 Testing"[Mesh]) OR ("COVID-19 vaccines"[Mesh]) OR ("COVID-19"[tiab]) OR ("Tertiary Care Centers"[Mesh]) OR ("Tertiary Care"[tiab]) OR ("Patient Care Team"[Mesh]) OR ("Patient Care Team"[tiab]) OR ("Expert center"[tiab]) OR ("Long-Term Care"[Mesh]) OR ("Long-Term Care"[tiab]) OR ("Interdisciplinary Communication"[MeSH]) OR ("Interdisciplinary"[tiab]) OR ("sequelae"[tiab]) OR ("associated disease"[tiab]) OR ("sequels"[tiab]) OR ("coexistent disease" [tiab]) OR ("concomitant disease" [tiab]) OR ("associated condition" [tiab]) OR ("coexistent condition" [tiab]) OR ("concomitant condition" [tiab]) OR ("Comorbidity"[MeSH]) OR ("Comorbidity"[tiab]) OR ("Myocardial Infarction"[Mesh]) OR ("Myocardial Infarction"[tiab]) OR ("Stroke"[Mesh]) OR ("Stroke"[tiab]) OR ("Staphylococcus aureus"[tiab]) OR ("Renal Insufficiency, Chronic"[Mesh]) OR ("Renal Insufficiency"[tiab]) OR ("renal function" [tiab]) OR ("renal failure" [tiab]) OR ("end-stage"[tiab]) AND ("kidney"[tiab]) OR ("renal"[tiab])) OR ("Pneumocystis"[Mesh]) OR ("Pneumocystis carinii"[Mesh]) OR ("Pneumocystis Infections"[Mesh]) OR ("Pneumonia, Pneumocystis"[Mesh]) OR ("Pneumocystis"[tiab]) OR ("Cardiovascular"[tiab]) OR ("prophyla**"[tiab]) OR ("Neoplasms" [Mesh]) OR ("Neoplasm" [tiab]) OR ("Cancer" [tiab]) OR ("malignancy" [tiab]) OR ("infectio*" [tiab]) OR ("Quality of Life" [Mesh]) OR ("Quality of Life" [tiab]))

AND

("2015/02/01"[Date - Publication] : "3000"[Date - Publication])

NOT

("Case reports" [Publication Type])
2.8. Search strategy General Management EMBASE

('ANCA associated vasculitis'/exp OR 'ANCA associated vasculitis*:ab,ti OR 'churg-strauss':ab,ti OR 'wegener:ab,ti OR ((granulomatosis:ab,ti OR microscopic:ab,ti) AND polyangiitis:ab,ti) OR ((ANCA:ab,ti OR 'neutrophil cytoplasmic antibody'/exp OR 'antineutrophil cytoplasmic antibody*:ab,ti) AND (vasculitis:ab,ti OR glomerulonephritis:ab,ti OR renal:ab,ti)) AND

('aged'/exp OR Aged:ab,ti OR Child*:ab,ti OR 'Child'/exp OR 'Adolescent'/exp OR Adolescent:ab,ti OR 'pregnancy'/exp OR pregnancy:ab,ti OR 'Contraceptive agent'/exp OR Contraception:ab,ti OR 'Kidney Transplantation'/exp OR 'Kidney Transplantation':ab,ti OR 'Antihypertensive Agent'/exp OR 'Antihypertensive therapy'/exp OR Antihypertensive:ab,ti OR 'Rehabilitation'/exp OR Rehabilitation:ab,ti OR 'patient care'/exp OR 'patient care':ab,ti OR 'shared decision making'/exp OR 'shared decision making':ab,ti OR 'Shared Medical Appointment*':ab,ti OR 'Patient Education'/exp OR 'Patient Education':ab,ti OR 'Risk Reduction'/exp OR 'Risk Reduction':ab,ti OR 'Patient-Reported Outcome'/exp OR 'Patient Reported Outcome':ab,ti OR 'fatigue'/exp OR Fatigue:ab,ti OR 'smoking'/exp OR Smoking:ab,ti OR 'vaccination'/exp OR Vaccination:ab,ti OR 'Severe acute respiratory syndrome coronavirus 2'/exp OR 'SARS-CoV-2':ab,ti OR 'coronavirus disease 2019'/exp OR 'sars-cov-2 vaccine'/exp OR 'COVID-19':ab,ti OR 'Tertiary Care Center'/exp OR 'Tertiary Care':ab,ti OR 'Patient Care Team':ab,ti OR 'Expert center':ab,ti OR 'Long-Term Care'/exp OR 'Long-Term Care':ab,ti OR 'Interdisciplinary Communication'/exp OR Interdisciplinary:ab,ti OR sequelae:ab,ti OR 'associated disease':ab,ti OR sequelae:ab,ti OR 'coexistent disease':ab,ti OR 'concomitant disease':ab,ti OR 'comorbidities':ab,ti OR 'coexistent condition':ab,ti OR 'concomitant condition':ab,ti OR 'heart infarction'/exp OR 'Myocardial Infarction':ab,ti OR 'cerebrovascular accident'/exp OR Stroke:ab,ti OR 'Staphylococcus aureus':ab,ti OR 'chronic kidney failure'/exp OR 'Renal Insufficiency':ab,ti OR 'renal function':ab,ti OR 'renal failure':ab,ti OR (end-stage:ab,ti AND (kidney:ab,ti OR renal:ab,ti)) OR 'Pneumocystis'/exp OR Pneumocystis:ab,ti OR Cardiovascular:ab,ti OR 'cardiovascular disease'/exp OR 'prophylaxis':ab,ti OR 'infection':ab,ti OR 'malignant neoplasm'/exp OR Neoplasm:ab,ti OR cancer:ab,ti OR malignancy:ab,ti OR 'quality of life'/exp OR 'quality of life':ab,ti)

AND

[01-02-2015]/sd

AND

([article]/lim OR [article in press]/lim OR [review]/lim)

NOT

'case report'/exp
2.9. Search strategy General Management Cochrane Library

ID Search
#1 MeSH descriptor: [Anti-Neutrophil Cytoplasmic Antibody-Associated Vasculitis] explode all trees
#2 (ANCA associated vasculiti* OR churg-strauss OR wegener):ti,ab,kw (Word variations have been searched)
#3 (granulomatosis OR microscopic):ti,ab,kw (Word variations have been searched)
#4 (polyangitis):ti,ab,kw (Word variations have been searched)
#5 #3 AND #4
#6 MeSH descriptor: [Antibodies, Antineutrophil Cytoplasmic] explode all trees
#7 (antineutrophil cytoplasmic antibod* OR ANCA):ti,ab,kw (Word variations have been searched)
#8 #6 OR #7
#9 (vasculitis OR glomerulonephritis OR renal):ti,ab,kw (Word variations have been searched)
#10 #8 AND #9
#11 #1 OR #2 OR #5 OR #10
#12 MeSH descriptor: [Aged] explode all trees
#13 MeSH descriptor: [Child] explode all trees
#14 MeSH descriptor: [Adolescent] explode all trees
#15 MeSH descriptor: [Contraception] explode all trees
#16 MeSH descriptor: [Kidney Transplantation] explode all trees
#17 MeSH descriptor: [Antihypertensive Agents] explode all trees
#18 MeSH descriptor: [Rehabilitation] explode all trees
#19 MeSH descriptor: [Patient Care] explode all trees
#20 MeSH descriptor: [Shared Medical Appointments] explode all trees
#21 MeSH descriptor: [Patient Education as Topic] explode all trees
#22 MeSH descriptor: [Risk Reduction Behavior] explode all trees
#23 MeSH descriptor: [Patient Reported Outcome Measures] explode all trees
#24 MeSH descriptor: [SARS-CoV-2] explode all trees
#25 MeSH descriptor: [COVID-19 Testing] explode all trees
#26 MeSH descriptor: [COVID-19 Vaccines] explode all trees
#27 MeSH descriptor: [Tertiary Care Centers] explode all trees
#28 MeSH descriptor: [Patient Care Team] explode all trees
#29 MeSH descriptor: [Long-Term Care] explode all trees
#30 MeSH descriptor: [Interdisciplinary Communication] explode all trees
#31 MeSH descriptor: [Myocardial Infarction] explode all trees
#32 MeSH descriptor: [Stroke] explode all trees
#33 MeSH descriptor: [Renal Insufficiency, Chronic] explode all trees
#34 MeSH descriptor: [Pneumocystis] explode all trees
#35 MeSH descriptor: [Pneumocystis carinii] explode all trees
#36 MeSH descriptor: [Pneumocystis infections] explode all trees
#37 MeSH descriptor: [Pneumonia, Pneumocystis] explode all trees
#38 MeSH descriptor: [Comorbidity] explode all trees
#39 MeSH descriptor: [Neoplasms] explode all trees
#40 MeSH descriptor: [Quality of Life] explode all trees
#41 (end-stage):ti,ab,kw (Word variations have been searched)
#42 (kidney OR renal):ti,ab,kw (Word variations have been searched)
#43 #41 AND #42
#44 (Aged OR Child* OR Adolescent OR pregnancy OR Contraception OR Kidney Transplantation OR Antihypertensive OR Rehabilitation OR patient care OR shared decision making OR Shared Medical Appointment* OR Patient Education OR Risk Reduction OR Patient Reported Outcome OR Fatigue OR Smoking OR Vaccination OR SARS-CoV-2 OR COVID-19 OR Tertiary Care OR Patient Care Team OR Expert center OR Long-Term Care OR Interdisciplinary OR sequelae OR associated disease OR sequels OR coexistent disease OR concomitant disease OR associated condition OR coexistent condition OR concomitant condition OR Myocardial Infarction OR Stroke OR Staphylococcus aureus OR Renal Insufficiency OR renal function OR renal failure OR end-stage AND kidney OR renal OR Pneumocystis OR Cardiovascular OR prophylax* OR comorbidit* OR neoplasm OR cancer OR malignancy OR infectio* OR quality of life):ti,ab,kw (Word variations have been searched)
#45 #12 OR #13 OR #14 OR #15 OR #16 OR #17 OR #18 OR #19 OR #20 OR #21 OR #22 OR #23 OR #24 OR #25 OR #26 OR #27 OR #28 OR #29 OR #30 OR #31 OR #32 OR #33 OR #34 OR #35 OR #36 OR #37 OR #38 OR #39 OR #40 OR #43 OR #44
#46 #11 AND #45 with Cochrane Library publication date Between Feb 2015 and Feb 2022
3. PRISMA diagrams

3.1. PRISMA diagram treatment (EGPA)

* 2 RCTs included in data synthesis for both GPA / MPA and EGPA.
Supplementary file 1: Treatment SLR EGPA, SLR Diagnostic & Follow-up, SLR General Management

3.2. PRISMA diagram diagnostic procedures and follow-up testing

[Diagram showing the PRISMA flowchart with PubMed, EMBASE, Cochrane Library, total records, duplicates removed, records screened, excluded by title/abstract screening, full-text articles assessed for eligibility, studies included, full-text articles excluded, with reasons: Total n = 95, Does not answer PICO (n = 67), Inclusion criteria not met (n = 24), Wrong article type (n = 4).]
3.3. PRISMA diagram general management

Supplementary file 1: Treatment SLR EGPA, SLR Diagnostic & Follow-up, SLR General Management

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