Supplementary ANCA-Associated Vasculitis Survey

Demographic Characteristics

1. **Do you spend ≥ 20% of your professional time in clinical practice?**
   ☐ Yes
   ☐ No
   If No selected: "This survey is intended for physicians who spend ≥ 20% of their professional time in clinical practice. You do not need to answer any further survey questions. Thank you for your time."

2. **What is your gender? (choose one)**
   ☐ Male
   ☐ Female

3. **What is your age? (specify in years)** ——————————

4. **What is your medical specialty? (choose one)**
   ☐ Rheumatology
   ☐ Pulmonology
   ☐ Nephrology
   ☐ Other – please specify——————————

5. **What is your level of training and years in practice? (choose one)**
   ☐ Fellow
   ☐ Attending physician, less than 5 years in practice
   ☐ Attending physician, 5-10 years in practice
   ☐ Attending physician, 11-20 years in practice
   ☐ Attending physician, >20 years in practice
   ☐ Other – please specify ——————————

8. **How many patients do you see per week? (choose one)**
   ☐ less than 5 per week
   ☐ 5-20 per week
   ☐ 21-50 per week
   ☐ more than 50 per week

9. **How many patients with granulomatosis with polyangiitis (Wegener's) or microscopic polyangiitis (ANCA-associated vasculidites) do you see per year? (choose one)**
   ☐ less than 5 per year
   ☐ 5-20 per year
   ☐ 21-50 per year
   ☐ more than 50 per year

Scenarios
Please read the following scenarios and choose the answer that best applies.

Scenario 1:
A patient with recurrent sinusitis, lung involvement (pulmonary nodules and ground glass opacities), positive serologies (anti-neurophil cytoplasmic antibody with anti-proteinase 3 antibody specificity by ELISA), renal involvement with active sediment including nephritic range proteinuria and red blood cell casts, normal creatinine, and biopsy-proven pauci-immune necrotising glomerulonephritis is diagnosed with ANCA-associated vasculitis. Infectious work-up is negative and the patient has normal liver function tests and a normal complete blood count (CBC). You are going to begin therapy.

1. **Along with corticosteroids, which of the following agents would you choose to treat this patient with if she were a <u>28 year old nulliparous female</u>? (choose one)**
   ☐ Oral daily cyclophosphamide (Cytoxan) – goal 2mg/kg/day
   ☐ Intravenous cyclophosphamide (Cytoxan) – 15mg/kg every 2 to 3 weeks
   ☐ Rituximab (Rituxan) – 375mg/m2 weekly for 4 weeks
   ☐ Methotrexate – target dose 20-25mg weekly
   ☐ Azathioprine (Imuran) – target dose 2mg/kg/day
   ☐ Mycopheolate mofetil (Cellcept) – target dose 2000-3000mg/day
   ☐ No preference
   ☐ Other – please specify——————————

2. **Please rate the importance of the following factors (efficacy, toxicity, cost/availability, comfort with use) with regard to how you made this decision. Provide a ranking on a scale of 1 to 4, with 1 being your first choice.**
   Efficacy data ——————————
   Toxicity data ——————————
   Cost/availability ——————————
   Comfort with use ——————————

3. **Is there another reason for your choice not listed above?**
   ☐ Yes
   If yes, please specify ——————————
   ☐ No

4. **Along with corticosteroids, which of the following agents would you choose to treat this patient with if she were a <u>68 year old female</u>? (choose one)**
   ☐ Oral daily cyclophosphamide (Cytoxan) – goal 2mg/kg/day
   ☐ Intravenous cyclophosphamide (Cytoxan) – 15mg/kg every 2 to 3 weeks
   ☐ Rituximab (Rituxan) – 375mg/m2 weekly for 4 weeks
   ☐ Methotrexate – target dose 20-25mg weekly
   ☐ Azathioprine (Imuran) – target dose 2mg/kg/day
   ☐ Mycopheolate mofetil (Cellcept) – target dose 2000-3000mg/day
   ☐ No preference
   ☐ Other – please specify——————————

5. **Please rate the importance of the following factors (efficacy, toxicity, cost/availability, comfort with use) with regard to how you made this decision. Provide a ranking on a scale of 1 to 4, with 1 being your first choice.**
   Efficacy data ——————————
   Toxicity data ——————————
   Cost/availability ——————————
   Comfort with use ——————————

6. **Is there another reason for your choice not listed above?**
   ☐ Yes
   If yes, please specify——————————
   ☐ No

7. **Along with corticosteroids, which of the following agents would you choose to treat this patient with if he were a <u>28 year old male</u>? (choose one)**
   ☐ Oral daily cyclophosphamide (Cytoxan) – goal 2mg/kg/day
   ☐ Intravenous cyclophosphamide (Cytoxan) – 15mg/kg every 2 to 3 weeks
   ☐ Rituximab (Rituxan) – 375mg/m2 weekly for 4 weeks
   ☐ Methotrexate – target dose 20-25mg weekly
   ☐ Azathioprine (Imuran) – target dose 2mg/kg/day
   ☐ Mycopheolate mofetil (Cellcept) – target dose 2000-3000mg/day
   ☐ No preference
   ☐ Other – please specify _____

8. **Please rate the importance of the following factors (efficacy, toxicity, cost/availability, comfort with use) with regard to how you made this decision. Provide a ranking on a scale of 1 to 4, with 1 being your first choice.**
   Efficacy data ————————————————
   Toxicity data ————————————————
   Cost/availability ———————————————
   Comfort with use ———————————————

9. **Is there another reason for your choice not listed above?**
   ☐ Yes
   If yes, please specify————————————
   ☐ No

10. **Along with corticosteroids, which of the following agents would you choose to treat this patient with if he were a <u>68 year old male</u>? (choose one)**
    ☐ Oral daily cyclophosphamide (Cytoxan) – goal 2mg/kg/day
    ☐ Intravenous cyclophosphamide (Cytoxan) – 15mg/kg every 2 to 3 weeks
    ☐ Rituximab (Rituxan) – 375mg/m2 weekly for 4 weeks
    ☐ Methotrexate – target dose 20-25mg weekly
    ☐ Azathioprine (Imuran) – target dose 2mg/kg/day
    ☐ Mycopheolate mofetil (Cellcept) – target dose 2000-3000mg/day
    ☐ No preference
    ☐ Other – please specify—————————————

11. **Please rate the importance of the following factors (efficacy, toxicity, cost/availability, comfort with use) with regard to how you made this decision. Provide a ranking on a scale of 1 to 4, with 1 being your first choice.**
    Efficacy data ————————————————
    Toxicity data ————————————————
    Cost/availability ———————————————
    Comfort with use ———————————————

12. **Is there another reason for your choice not listed above?**
    ☐ Yes
    If yes, please specify————————————
    ☐ No

Scenario 2:
The patient from scenario 1 (see attachment below) undergoes induction therapy. At 4 months symptoms, laboratory values and urine normalise. The patient takes prednisone 10mg daily, which is being tapered.

[Attachment: "Scenario 1.doc"]

1. **Which of the following agents would you choose to continue or start on this patient at this time if she were a <u>28 year old nulliparous female</u>? (choose one)**
   ☐ Oral daily cyclophosphamide (Cytoxan)
   ☐ Intravenous cyclophosphamide (Cytoxan)
   ☐ Rituximab (Rituxan)
   ☐ Methotrexate
   ☐ Azathioprine (Imuran)
   ☐ Mycopheolate mofetil (Cellcept)
   ☐ Trimethoprim/sulfamethoxazole 160/800 (Bactrim DS) – one tablet twice daily
   ☐ Leflunomide (Arava)
   ☐ No additional immunosuppressant therapy. Follow expectantly.
   ☐ No preference
   ☐ Other – please specify—————————————

2. **Please rate the importance of the following factors (efficacy, toxicity, cost/availability, comfort with use) with regard to how you made this decision. Provide a ranking on a scale of 1 to 4, with 1 being your first choice.**
   Efficacy data ————————————————
   Toxicity data ————————————————
   Cost/availability ———————————————
   Comfort with use ———————————————

3. **Is there another reason for your choice not listed above?**
   ☐ Yes
   If yes, please specify————————————
   ☐ No

4. **Along with corticosteroids, which of the following agents would you choose to continue or start on this patient at this time if she were a <u>68 year old female</u>? (choose one)**
   ☐ Oral daily cyclophosphamide (Cytoxan)
   ☐ Intravenous cyclophosphamide (Cytoxan)
   ☐ Rituximab (Rituxan)
   ☐ Methotrexate
   ☐ Azathioprine (Imuran)
   ☐ Mycopheolate mofetil (Cellcept)
   ☐ Trimethoprim/sulfamethoxazole 160/800 (Bactrim DS) – one tablet twice daily
   ☐ Leflunomide (Arava)
   ☐ No additional immunosuppressant therapy. Follow expectantly.
   ☐ No preference
   ☐ Other – please specify—————————————

5. **Please rate the importance of the following factors (efficacy, toxicity, cost/availability, comfort with use) with regard to how you made this decision. Provide a ranking on a scale of 1 to 4, with 1 being your first choice.**
   Efficacy data ————————————————
   Toxicity data ————————————————
   Cost/availability ———————————————
   Comfort with use ———————————————

6. **Is there another reason for your choice not listed above?**
   ☐ Yes
   If yes, please specify ———————————
   ☐ No

7. **Along with corticosteroids, which of the following agents would you choose to continue or start on this patient at this time if he were a 28 year old male? (choose one)**
   ☐ Oral daily cyclophosphamide (Cytoxan)
   ☐ Intravenous cyclophosphamide (Cytoxan)
   ☐ Rituximab (Rituxan)
   ☐ Methotrexate
   ☐ Azathioprine (Imuran)
   ☐ Mycopheolate mofetil (Cellcept)
   ☐ Trimethoprim/sulfamethoxazole 160/800 (Bactrim DS) – one tablet twice daily
   ☐ Leflunomide (Arava)
   ☐ No additional immunosuppressant therapy. Follow expectantly.
   ☐ No preference
   ☐ Other – please specify——————————————

8. **Please rate the importance of the following factors (efficacy, toxicity, cost/availability, comfort with use) with regard to how you made this decision. Provide a ranking on a scale of 1 to 4, with 1 being your first choice.**
   Efficacy data ——————————————————
   Toxicity data ——————————————————
   Cost/availability ————————————————
   Comfort with use ————————————————

9. **Is there another reason for your choice not listed above?**
   ☐ Yes
   If yes, please specify ———————————————
   ☐ No

10. **Along with corticosteroids, which of the following agents would you choose to continue or start on this patient at this time if he were a 68 year old male? (choose one)**
    ☐ Oral daily cyclophosphamide (Cytoxan)
    ☐ Intravenous cyclophosphamide (Cytoxan)
    ☐ Rituximab (Rituxan)
    ☐ Methotrexate
    ☐ Azathioprine (Imuran)
    ☐ Mycopheolate mofetil (Cellcept)
    ☐ Trimethoprim/sulfamethoxazole 160/800 (Bactrim DS) – one tablet twice daily
    ☐ Leflunomide (Arava)
    ☐ No additional immunosuppressant therapy. Follow expectantly.
    ☐ No preference
    ☐ Other – please specify——————————————

11. **Please rate the importance of the following factors (efficacy, toxicity, cost/availability, comfort with use) with regard to how you made this decision. Provide a ranking on a scale of 1 to 4, with 1 being your first choice.**
    Efficacy data ——————————————————
    Toxicity data ——————————————————
    Cost/availability ————————————————
    Comfort with use ————————————————

12. **Is there another reason for your choice not listed above?**
    ☐ Yes
    If yes, please specify ———————————————
    ☐ No

Scenario 3:
A patient with no history of cocaine use has a saddle nose deformity, active sinusitis, positive serologies (anti-neutrophil cytoplasmic antibody with anti-proteinase 3 antibody specificity by ELISA), no renal involvement, tracheal narrowing and multiple small pulmonary nodules on computerised tomography. A biopsy of one of these nodules by video-assisted thoracic surgery (VATS) shows necrotising granulomatous inflammation with pauci-immune vasculitis, and the patient is diagnosed with ANCA-associated vasculitis. The infectious work-up is negative and the patient has normal liver function tests and a normal complete blood count (CBC).

1. **Along with corticosteroids, which of the following agents would you choose to start on this patient at this time if she were a 28 year old nulliparous female? (choose one)**
   ☐ Oral daily cyclophosphamide (Cytoxan)
   ☐ Intravenous cyclophosphamide (Cytoxan)
   ☐ Rituximab (Rituxan)
   ☐ Methotrexate
   ☐ Azathioprine (Imuran)
   ☐ Mycopheolate mofetil (Cellcept)
   ☐ Trimethoprim/sulfamethoxazole 160/800 (Bactrim DS) – one tablet twice daily
   ☐ Leflunomide (Arava)
   ☐ No preference
   ☐ Other – please specify——————————————

2. **Please rate the importance of the following factors (efficacy, toxicity, cost/availability, comfort with use) with regard to how you made this decision. Provide a ranking on a scale of 1 to 4, with 1 being your first choice.**
   Efficacy data ——————————————————
   Toxicity data ——————————————————
   Cost/availability ————————————————
   Comfort with use ————————————————

3. **Is there another reason for your choice not listed above?**
   ☐ Yes
   If yes, please specify ———————————————
   ☐ No

4. **Along with corticosteroids, which of the following agents would you choose to start on this patient at this time if she were a 68 year old female? (choose one)**
   ☐ Oral daily cyclophosphamide (Cytoxan)
   ☐ Intravenous cyclophosphamide (Cytoxan)
   ☐ Rituximab (Rituxan)
   ☐ Methotrexate
   ☐ Azathioprine (Imuran)
   ☐ Mycopheolate mofetil (Cellcept)
   ☐ Trimethoprim/sulfamethoxazole 160/800 (Bactrim DS) – one tablet twice daily
   ☐ Leflunomide (Arava)
   ☐ No preference
   ☐ Other – please specify——————————————

5. **Please rate the importance of the following factors (efficacy, toxicity, cost/availability, comfort with use) with regard to how you made this decision. Provide a ranking on a scale of 1 to 4, with 1 being your first choice.**
   Efficacy data ——————————————————
   Toxicity data ——————————————————

Cost/availability ——————————————————
Comfort with use ——————————————————

6. **Is there another reason for your choice not listed above?**
   ☐ Yes
   If yes, please specify——————————————
   ☐ No

7. **Along with corticosteroids, which of the following agents would you choose to start on this patient at this patient if he were a 28 year old male? (choose one)**
   ☐ Oral daily cyclophosphamide (Cytoxan)
   ☐ Intravenous cyclophosphamide (Cytoxan)
   ☐ Rituximab (Rituxan)
   ☐ Methotrexate
   ☐ Azathioprine (Imuran)
   ☐ Mycopheolate mofetil (Cellcept)
   ☐ Trimethoprim/sulfamethoxazole 160/800 (Bactrim DS) – one tablet twice daily
   ☐ Leflunomide (Arava)
   ☐ No preference
   ☐ Other – please specify ————————————

8. **Please rate the importance of the following factors (efficacy, toxicity, cost/availability, comfort with use) with regard to how you made this decision. Provide a ranking on a scale of 1 to 4, with 1 being your first choice.**
   Efficacy data ——————————————————
   Toxicity data ——————————————————
   Cost/availability ——————————————————
   Comfort with use ——————————————————

9. **Is there another reason for your choice not listed above?**
   ☐ Yes
   If yes, please specify——————————————
   ☐ No

10. **Along with corticosteroids, which of the following agents would you choose to start on this patient if he were a 68 year old male? (choose one)**
    ☐ Oral daily cyclophosphamide (Cytoxan)
    ☐ Intravenous cyclophosphamide (Cytoxan)
    ☐ Rituximab (Rituxan)
    ☐ Methotrexate
    ☐ Azathioprine (Imuran)
    ☐ Mycopheolate mofetil (Cellcept)
    ☐ Trimethoprim/sulfamethoxazole 160/800 (Bactrim DS) – one tablet twice daily
    ☐ Leflunomide (Arava)
    ☐ No preference
    ☐ Other – please specify——————————

11. **Please rate the importance of the following factors (efficacy, toxicity, cost/availability, comfort with use) with regard to how you made this decision. Provide a ranking on a scale of 1 to 4, with 1 being your first choice.**
    Efficacy data ——————————————————
    Toxicity data ——————————————————
    Cost/availability ——————————————————
    Comfort with use ——————————————————

12. **Is there another reason for your choice not listed above?**
    ☐ Yes
    If yes, please specify ————————————
    ☐ No

Thank you for your participation.