

Supplementary File 1. Pediatric survey.

Relapsing Polychondritis Epidemiology in Children

We are asking you to volunteer to take part in a survey as part of a research study about the epidemiology of relapsing polychondritis (RP) in children. This survey is being conducted to learn more about the clinical presentations, triggers and complications of relapsing polychondritis in patients less than 18 years old. A parent or relative taking care of the subject can complete the survey. If the child is older than 16, he or she may complete the survey. Your participation in this survey is completely voluntary. This means you do not have to participate if you don't want to. If you agree to participate, you have the right to only answer the questions you choose to answer. We will not collect any personally identifying information about you. This information may be shared with members of the research team and the relapsing polychondritis foundation.

1. Who is answering the survey?

- ☐ The patient (only for patients older than 16 years old)
- ☐ Mother
- ☐ Father
- ☐ Sibling
- ☐ Other

2. The patient is?

- ☐ Female
- ☐ Male

3. How old is the patient? (years)

4. The patient is?

- ☐ White
- ☐ African American
- ☐ American Indian or Alaskan
- ☐ Asian
- ☐ Hawaiian or Pacific Islander
- ☐ Hispanic
- ☐ More than once race
- ☐ Other

5. At what age (years) did the patient start first presenting symptoms attributable to Relapsing Polychondritis?

6. What is the patient's country of origin?

7. What country does the patient currently live in?

8. How old was the patient when he/she was first diagnosed with Relapsing Polychondritis?

9. Who diagnosed the patient?

- ☐ Primary care doctor
- ☐ Rheumatologist
- ☐ ENT
- ☐ Other (please specify): _____

10. How many doctors did the patient need to see before a diagnosis was made?

- ☐ One doctor
- ☐ Two doctors
- ☐ Three doctors
- ☐ More than three

11. Did the patient undergo biopsy in order to support the diagnosis?

- ☐ Yes
- ☐ No
- ☐ If yes, please specify the location (i.e. trachea, ear): _____

12. What were the patient's initial symptoms before diagnosis? (please check all that apply)

- ☐ Nose pain/pressure
- ☐ Nose redness
- ☐ Ear pain
- ☐ Ear redness
- ☐ Changes in your voice/hoarseness
- ☐ Shortness of breath
- ☐ Breathing pain
- ☐ Knee pain
- ☐ Ankle pain
- ☐ Elbow pain
- ☐ Wrist pain
- ☐ Finger pain
- ☐ Knee swelling
- ☐ Ankle swelling
- ☐ Elbow swelling
- ☐ Wrist swelling
- ☐ Finger swelling
- ☐ Other joint pain (spine, shoulder, mandibular joint, pelvic joints)
- ☐ Costochondritis (chest wall pain)
- ☐ Dizziness
- ☐ Hearing loss

- ☐ Eye inflammation (episcleritis, iritis, corneal ulcer, uveitis)
- ☐ Raynaud's (purple color on hands and/or feet with cold exposure)
- ☐ Difficulty Swallowing
- ☐ Other (please specify): _____

13. Please check all the patient's current symptoms?

- ☐ Nose pain/pressure
- ☐ Nose redness
- ☐ Ear pain
- ☐ Ear redness
- ☐ Changes in your voice/hoarseness
- ☐ Shortness of breath
- ☐ Breathing pain
- ☐ Knee pain
- ☐ Ankle pain
- ☐ Elbow pain
- ☐ Wrist pain
- ☐ Finger pain
- ☐ Knee swelling
- ☐ Ankle swelling
- ☐ Elbow swelling
- ☐ Wrist swelling
- ☐ Finger swelling
- ☐ Other joint pain (spine, shoulder, mandibular joint, pelvic joints)
- ☐ Costochondritis
- ☐ Dizziness
- ☐ Hearing loss
- ☐ Eye inflammation (episcleritis, iritis, corneal ulcer, uveitis)
- ☐ Raynaud's (purple color on hands and/or feet with cold exposure)
- ☐ Difficulty swallowing
- ☐ Other (please specify): _____

14. How many years after the patient started to have symptoms was the diagnosis of RP made?

- ☐ Less than a year
- ☐ 1-3 years
- ☐ 3-5 years
- ☐ 5-8 years
- ☐ 8-10 years
- ☐ More than 10 years

15. Did the patient go to the emergency room due to RP related symptoms before a diagnosis was made?

- ☐ Yes
- ☐ No

16. If yes, how many times did the patient go to the emergency room due to symptoms related to RP before a diagnosis was made?

- ☐ one time
- ☐ two times
- ☐ three times
- ☐ four times
- ☐ five times
- ☐ more than five times

17. Please check the RP symptoms that prompted the patient to go to the ER. Check all that apply

- ☐ Shortness of breath
- ☐ Chest pain/costochondritis
- ☐ Throat pain
- ☐ Difficulty swallowing
- ☐ Dizziness
- ☐ Ear pain
- ☐ Nose pain
- ☐ Eye pain
- ☐ Pain when you breathe
- ☐ Other (please specify): _____

18. Was the patient diagnosed with sinusitis before RP was diagnosed?

- ☐ Yes
- ☐ No
- ☐ If yes, how long before you were diagnosed with RP?: _____

19. Was the patient diagnosed with asthma before RP was diagnosed?

- ☐ Yes
- ☐ No
- ☐ If yes, how long before you were diagnosed with RP?: _____

20. Was the patient diagnosed with repeated ear infections before RP was diagnosed?

- ☐ Yes
- ☐ No
- ☐ If yes, how many months/years before you were diagnosed with RP?: _____

21. Is there another blood relative with confirmed diagnosis of RP?

- ☐ Brother
- ☐ Sister
- ☐ Mother
- ☐ Father
- ☐ Not applicable
- ☐ Other (please specify): _____

22. Does the patient have any other autoimmune disease?

- ☐ Yes
- ☐ No

23. Please check all the autoimmune diseases that apply

- ☐ Not applicable
- ☐ Arthritis
- ☐ Lupus erythematosus
- ☐ Vasculitis
- ☐ Thyroiditis
- ☐ Common variable immunodeficiency
- ☐ Vitiligo
- ☐ Diabetes mellitus type I
- ☐ Other (please specify): _____

24. Was the other autoimmune disease diagnosed before RP?

- ☐ Yes
- ☐ No
- ☐ How long before RP diagnosis? _____

25. Does the patient have any other medical conditions? (please check all that apply)

- ☐ Ulcerative colitis
- ☐ Crohn's disease
- ☐ Myelodysplastic syndrome
- ☐ Acute myelogenous leukemia
- ☐ Renal failure

26. Does the patient have worsening of her/his symptoms 24 hours after eating any of the following foods?

- ☐ No worsening of symptoms related to food
- ☐ Gluten
- ☐ Soy
- ☐ Red meat
- ☐ High sugar content food
- ☐ High fat content food
- ☐ If more than one food listed above, or other food not listed worsens your symptoms, please specify here: _____

27. Does the patient have worsening of her/his symptoms 24-48 hours after any of the following?

- ☐ Exhausting Physical activity
- ☐ Lack of sleep
- ☐ Change in weather
- ☐ Bronchoscopy
- ☐ Surgery
- ☐ Piercing
- ☐ Airplane travel
- ☐ Emotional stressful situations
- ☐ Sun exposure
- ☐ Physical injury

- ☐ If more than one activity listed above, or other not listed worsens your symptoms, please specify here: _____

28. Has the patient had a flare after any of the following infections? (please check all that apply)

- ☐ Any infection
☐ Cold or upper respiratory infection
☐ Gastroenteritis
☐ Urinary tract infection
☐ Strep throat
☐ Pneumonia
☐ Other (please specify): _____

29. For girls only

Does the patient notice worsening, or improving, of her symptoms around the time of her menstrual cycle?

- ☐ Worsening of the symptoms
☐ Improving of the symptoms
☐ Not applicable

30. Is the patient currently taking steroids?

- ☐ Yes
☐ No
☐ What is the name of the steroid?: _____
☐ How many milligrams?: _____
☐ For how long?: _____

31. Which medications does the patient take for RP now? (check all that apply)

- ☐ Methotrexate
☐ Cyclophosphamide/Cytosan
☐ Azathioprine (Imuran)
☐ Dapsone
☐ Prednisone
☐ Tocilizumab (Actemra)
☐ Anakinra
☐ Remicade
☐ Enbrel
☐ Humira
☐ Other (please specify): _____

32. Which medications has the patient taken in the past for RP before their current regimen? (check all that apply)

- ☐ Methotrexate
☐ Cyclophosphamide
☐ Azathioprine
☐ Prednisone
☐ Methotrexate and prednisone

- ☐ Cyclophosphamide and prednisone
- ☐ Azathioprine and prednisone
- ☐ Tocilizumab (Actemra)
- ☐ Anakinra
- ☐ Enbrel
- ☐ Humira
- ☐ Remicade
- ☐ Other (please specify): _____

33. What is the highest dose of prednisone that the patient has to take to control an acute flare?

34. How many flares did the patient get per month prior to diagnosis?

- ☐ one
- ☐ two
- ☐ three
- ☐ more than three

35. How many flares does the patient get per month since he/she started treatment?

- ☐ One
- ☐ Two
- ☐ Three
- ☐ More than three

36. How long has the longest flare lasted?

37. Has the patient been diagnosed with tracheomalacia/weakness/collapse of the trachea?

- ☐ Yes
- ☐ No
- ☐ If you answer yes, have you had a tracheostomy?: _____

38. Has the patient had any of the following tests done? (please include all that apply)

- ☐ Echocardiogram
- ☐ Pulmonary function test
- ☐ CT scan of the chest
- ☐ CT scan of the neck
- ☐ Bronchoscopy
- ☐ Audiology evaluation
- ☐ Ophthalmology evaluation
- ☐ Other (please specify): _____

39. Has the patient had pneumonia due to RP?

- ☐ Yes

- ☐ No
- ☐ If you answer yes, how many times?: _____

40. Has the patient been intubated due to RP?

- ☐ Yes
- ☐ No
- ☐ If you answer yes, please specify why (infection/RP/inflammation of your bronchial tree):

41. How many times has the patient been hospitalized due to RP?

42. Has RP interrupted the patient's education for more than a week at a time?

- ☐ Yes
- ☐ No

43. Has RP interrupted the patient's education for more than a month at a time?

- ☐ Yes
- ☐ No

44. Does the patient take any of the following medications? please check all that apply

- ☐ Medication to control blood pressure
- ☐ Medication to control high blood sugar
- ☐ Medication to control cholesterol
- ☐ Eye drop medications
- ☐ Stomach protecting medication
- ☐ Medication to control arthritis

45. Does the patient use a hearing aid?

- ☐ Yes
- ☐ No

46. Does the patient need any of the following?

- ☐ Wheelchair
- ☐ Cane
- ☐ None
- ☐ Other (please specify): _____

47. Who is/are the current doctor(s) that help manage the patient's RP (check all that apply)

- ☐ ENT
- ☐ Rheumatologist
- ☐ Primary care
- ☐ Pulmonologist
- ☐ Cardiologist

Supplementary File 2. Adult survey.

Adult Relapsing Polychondritis Epidemiology

Thank you so much for spending the time filling this survey! You are making a difference and helping tremendously other patients with RP! Your participation in this survey is completely voluntary. This means you do not have to participate if you don't want to. If you agree to participate, you have the right to only answer the questions you choose to answer. We will not collect any personally identifying information about you. This information may be shared with members of the research team and the relapsing polychondritis foundation.

1. Are you

- ☐ Female
- ☐ Male

2. How old are you?

3. Are you?

- ☐ White
- ☐ African American
- ☐ Indian or Alaskan
- ☐ Asian
- ☐ Hawaiian or Pacific Islander
- ☐ Hispanic
- ☐ More than once race

4. What is your country of origin?

5. When were you diagnosed with RP?

Date / Time: _____

6. Who diagnosed you?

- ☐ Primary care doctor
- ☐ Rheumatologist
- ☐ ENT
- ☐ Other (please specify): _____

7. How many doctors did you see before you were diagnosed with RP?

- ☐ One doctor
- ☐ Two doctors
- ☐ Three doctors
- ☐ More than three
- ☐

8. Did you undergo a cartilage biopsy in order to support your diagnosis?

- ☐ Yes
- ☐ No

9. What were your primary symptoms before diagnosis (please check all that apply)

- ☐ Nose pain/pressure
- ☐ Nose redness
- ☐ Ear pain
- ☐ Changes in your voice/hoarseness
- ☐ Shortness of breath
- ☐ Breathing pain
- ☐ Knee pain
- ☐ Ankle pain
- ☐ Elbow pain
- ☐ Wrist pain
- ☐ Finger pain
- ☐ Knee swelling
- ☐ Ankle swelling
- ☐ Elbow swelling
- ☐ Wrist swelling
- ☐ Finger swelling
- ☐ Other joint pain (spine, shoulder, mandibular joint, pelvic joints)
- ☐ Costochondritis
- ☐ Dizziness
- ☐ Hearing loss
- ☐ Eye inflammation (episcleritis, iritis, corneal ulcer, uveitis)
- ☐ Raynaud's (purple color on hands and/or feet with cold exposure)
- ☐ Other (please specify): _____

10. Please check all the symptoms you had before and after diagnosis (please check all that apply)

- ☐ Nose pain/pressure
- ☐ Nose redness
- ☐ Ear pain
- ☐ Changes in your voice/hoarseness
- ☐ Shortness of breath
- ☐ Breathing pain
- ☐ Knee pain
- ☐ Ankle pain
- ☐ Elbow pain
- ☐ Wrist pain
- ☐ Finger pain
- ☐ Knee swelling
- ☐ Ankle swelling
- ☐ Elbow swelling
- ☐ Wrist swelling
- ☐ Finger swelling
- ☐ Other joint pain (spine, shoulder, mandibular joint, pelvic joints)

- ☐ Costochondritis
- ☐ Dizziness
- ☐ Hearing loss
- ☐ Eye inflammation (episcleritis, iritis, corneal ulcer, uveitis)
- ☐ Raynaud's (purple color on hands and/or feet with cold exposure)
- ☐ Other (please specify): _____

11. How many years after you started to have symptoms were you diagnosed with RP?

- ☐ less than a year
- ☐ 1-3 years
- ☐ 3-5 years
- ☐ 5-8 years
- ☐ 8-10 years
- ☐ more than 10 years

12. Did you go to the emergency room due to RP related symptoms before you had a diagnosis?

- ☐ Yes
- ☐ No

13. How many times did you go to the ER?

- ☐ one time
- ☐ two times
- ☐ three times
- ☐ four times
- ☐ five times
- ☐ more than five times

14. Please check the symptoms that prompted you to go to the ER. Check all that apply:

- ☐ Shortness of breath
- ☐ Chest pain/costochondritis
- ☐ Throat pain
- ☐ Difficulty swallowing
- ☐ Dizziness
- ☐ Ear pain
- ☐ Nose pain
- ☐ Eye pain
- ☐ Pain when you breathe
- ☐ Other (please specify): _____

15. Were you diagnosed with sinusitis before you were diagnosed with RP?

- ☐ Yes
- ☐ No
- ☐ If you answer yes, how long before you were diagnosed with RP: _____

16. Were you diagnosed with asthma before you were diagnosed with RP?

- ☐ Yes
- ☐ No
- ☐ If you answer yes, how long before you were diagnosed with RP?: _____

17. Were you diagnosed with muscular tension dysphonia before you were diagnosed with RP?

- ☐ Yes
- ☐ No
- ☐ If you answer yes, how many months/years before you were diagnosed with RP?: _____

18. Were you diagnosed with fibromyalgia before you were diagnosed with RP?

- ☐ Yes
- ☐ No
- ☐ If you answer yes, how many months/years before you were diagnosed with RP?: _____

19. Were you diagnosed with an ear infection before you were diagnosed with RP?

- ☐ Yes
- ☐ No
- ☐ If you answer yes, how many months/years before you were diagnosed with RP?: _____

20. Is there another blood relative member with a confirmed diagnosis of RP?

- ☐ Brother
- ☐ Sister
- ☐ Mother
- ☐ Father
- ☐ Not applicable
- ☐ Other (please specify): _____

21. Do you have any other autoimmune disease?

- ☐ Yes
- ☐ No

22. Please check all that apply

- ☐ Not applicable
- ☐ Rheumatoid arthritis
- ☐ Lupus erythematosus
- ☐ Vasculitis
- ☐ Thyroiditis
- ☐ Common variable immunodeficiency
- ☐ Other (please specify): _____

23. Was the other autoimmune disease diagnosed before RP?

- ☐ Yes
- ☐ No
- ☐ How long before you had the diagnosis of RP?: _____

24. Do you notice worsening of your symptoms 24 hours after eating any of the following foods?

- ☐ I do not have any worsening of my symptoms related to food
- ☐ Gluten
- ☐ Soy
- ☐ Red meat
- ☐ High sugar content food
- ☐ High fat content food
- ☐ Alcohol
- ☐ If more than one food listed above, or other food not listed worsens your symptoms, please specify here: _____

25. Do you notice worsening of your symptoms 24-48 hours after any of the following?

- ☐ Exhausting Physical activity
- ☐ Lack of sleep
- ☐ Change in weather
- ☐ Bronchoscopy
- ☐ Surgery
- ☐ Piercing
- ☐ Airplane travel
- ☐ If more than one activity listed above, or other not listed worsens your symptoms, please specify here: _____

26. Have you had a flare after any of the following infections? (please check all that apply)

- ☐ Any infection
- ☐ Cold or upper respiratory infection
- ☐ Gastroenteritis
- ☐ Urinary tract infection
- ☐ Strep throat
- ☐ Pneumonia
- ☐ Other (please specify): _____

27. Have you had a flare after any of the following situations? (please check all that apply)

- ☐ Change in school or Job
- ☐ Serious financial difficulties
- ☐ Spouse, partner or close friend death
- ☐ Move to a new house
- ☐ Loss of Job
- ☐ Separation or divorce
- ☐ Other (please specify): _____

28. Did you have a flare after pregnancy?

- ☐ Yes
- ☐ No

- ☐ Not applicable
- ☐ If yes, how long after delivery?: _____

29. Do you notice any change in your symptoms when you get your period?

- ☐ Worsening of the symptoms
- ☐ Improving of the symptoms
- ☐ Not applicable

30. Which medications do you take for RP now?

- ☐ Methotrexate
- ☐ Cyclophosphamide/Cytosan
- ☐ Azathioprine (Imuran)
- ☐ Dapsone
- ☐ Prednisone
- ☐ Other (please specify): _____

31. Which medications have you taken in the past for RP before your current regimen?

- ☐ Methotrexate
- ☐ Cyclophosphamide
- ☐ Azathioprine
- ☐ Prednisone
- ☐ Methotrexate and prednisone
- ☐ Cyclophosphamide and prednisone
- ☐ Azathioprine and prednisone
- ☐ Other (please specify): _____

32. What is the highest dose of prednisone that you have taken to control your disease?

33. How many flares did you get per month prior to your diagnosis?

- ☐ one
- ☐ two
- ☐ three
- ☐ more than three

34. How many flares do you get per month since you started treatment?

- ☐ One
- ☐ Two
- ☐ Three
- ☐ More than three

35. How long has the longest flare lasted?

36. Have you been diagnosed with tracheomalacia/weakness/collapse of the trachea?

- ☐ Yes
- ☐ No
- ☐ If you answer yes, have you had a tracheostomy?: _____

37. Have you had any of the following test done?

- ☐ Echocardiogram
- ☐ Pulmonary function test
- ☐ CT scan of the chest
- ☐ CT scan of the neck
- ☐ Bronchoscopy
- ☐ Audiology evaluation
- ☐ Ophthalmology evaluation
- ☐ Other (please specify)

38. Have you had pneumonia after your diagnosis of RP?

- ☐ Yes
- ☐ No
- ☐ If you answer yes, how many times?

39. Have you been intubated?

- ☐ Yes
- ☐ No
- ☐ If you answer yes, please specify why (infection/RP/inflammation of your bronchial tree):

40. Are you on disability due to RP?

- ☐ Yes
- ☐ No

41. Why did you have to go on disability?

42. How long after you were diagnosed with RP did you stop working?

43. Did you undergo body piercing before you were diagnosed with RP?

- ☐ Yes
- ☐ No
- ☐ If you answer yes, which body part and how long prior your diagnosis?: _____

44. Do you rely upon alternative medicine/complimentary medicine interventions for management of your disease?

- ☐ Yes
- ☐ No

☐ If you answer yes, please specify treatment: _____

45. Do you manage your symptoms by adhering to any particular dietary modifications?

- ☐ Yes
☐ No
☐ If you answer yes, please specify: _____

46. Have you ever smoked?

- ☐ Yes
☐ No

47. If yes, how many years have you smoked?

48. How many packs per day?

Supplementary Table 1. Comparison of patient-reported joint complaints between patients with an adult or a pediatric diagnosis of relapsing polychondritis.

	Adult (n=290)	Pediatric (n=21)	<i>p</i>-value
Knee Pain/Swelling	168 (57.9%)	16 (76.2%)	$p = 0.11$
Ankle Pain/Swelling	147 (50.7%)	14 (66.7%)	$p = 0.18$
Elbow Pain/Swelling	113 (39.0%)	4 (19.0%)	$p = 0.10$
Finger Pain/Swelling	171 (59.0%)	8 (38.1%)	$p = 0.07$
Wrist Pain/Swelling	151 (52.1%)	9 (42.9%)	$p = 0.50$
Other Joint Pain/Swelling	194 (66.9%)	13 (61.9%)	$p = 0.64$
Any Joint Pain/Swelling	248 (85.5%)	19 (90.5%)	$p = 0.75$