CRYOPYRIN ASSOCIATED PERIODIC SYNDROME (CAPS)
Australian Paediatric Surveillance Unit

Please contact Dr SAM MEHR on (02) 9845-3420 or samm@chw.edu.au or the APSU on (02) 9845-3005 or apsu@chw.edu.au if you have any questions about this form.

Instructions: Please complete a questionnaire for each patient under your care.
Please answer each question by ticking the appropriate box or writing your response in the space provided. DK=Don't Know

REPORTING CLINICIANS DETAILS:

1. APSU Dr Code/Name: ☐☐☐☐ /____________________________

2. Date questionnaire completed: ☐☐ / ☐☐ / ☐☐

3. Specialist position: ☐ General Paediatrician ☐ Dermatologist ☐ Immunologist ☐ Neurologist ☐ Rheumatologist ☐ Other (please specify)______________________________

PATIENT DETAILS:

4. First 2 letters of first name: ☐☐ Surname: ☐☐ **5.** Date of Birth: ☐☐ / ☐☐ / ☐☐

6. Gender: ☐ M ☐ F **7.** Postcode: ☐☐☐☐

8. Date of diagnosis: ☐☐ / ☐☐ / ☐☐ (if not known, then age of diagnosis ________(in yrs) or____(in mos)

9. Country of birth: ☐ Australia ☐ Other specify____

10. Child's ethnicity: ☐ Aboriginal ☐ Torres Strait Islander ☐ Caucasian ☐ Pacific Islander ☐ Maori ☐ Asian ☐ Middle Eastern ☐ African ☐ Other: Please specify:__________________ ☐ DK

11a. Mother's country of birth: ☐ Australia ☐ Other specify:________________________________ ☐ DK

11b. Father's country of birth: ☐ Australia ☐ Other specify:________________________________ ☐ DK

If this patient is primarily cared for by another physician who you believe will report the case and could provide additional details, please write the other physician's name and hospital below, complete questionnaire details above this line and return to APSU. If no other report is received for this child we will contact you for further information requested in the remainder of the questionnaire.

The primary clinician caring for this child is: ***Name:*** ***Hospital:***

12. SPECIFY TYPE OF CRYOPYRIN ASSOCIATED PERIODIC FEVER SYNDROME (CAPS)

a. NOMID/CINCA ☐ Yes ☐ No
b. Familial Cold Auto-inflammatory Syndrome (FCAS) ☐ Yes ☐ No
c. Muckle Wells Syndrome (MWS) ☐ Yes ☐ No
d. Suspect CAPS but type unknown ☐ Yes ☐ No

13. GENE MUTATIONAL ANALYSIS PERFORMED (CIAS1/NLRP3 GENE)

a. ☐ Not performed ☐ Performed but no mutation found ☐ Performed and mutation found ☐ DK
b. If mutation found, please specify mutation detected:____
c. Parent(s) found to be carriers: ☐ Yes ☐ No ☐ Not performed; *If YES* which parent(s):______________

CLINICAL FEATURES PRESENT AT ANY STAGE IN THE ABSENCE OF ANTI-IL-1 THERAPY

14. Approximate date of first presentation: ☐☐ / ☐☐ / ☐☐ or age_____(yrs) ______(months)

15. Urticarial like rash ☐ Yes ☐ No ☐ DK
a. Age (months) of onset of urticarial like rash:_______________months
b. Frequency of urticarial like rash: ☐ daily ☐ weekly ☐ monthly ☐ Other (Specify):_____________ ☐ DK
c. Biopsy of rash performed? ☐ Yes ☐ No ☐ DK If YES, Pathology conclusion:____________________

16. Neurological/CNS feature(s)
a. Chronic headaches (at least 15 headaches/month for ≥ 3 months) ☐ Yes ☐ No ☐ DK
b. Aseptic meningitis on lumbar puncture ☐ Yes ☐ No ☐ DK
c. Macrocephaly (head circumference > 95% centile) ☐ Yes ☐ No ☐ DK
d. Developmental delay ☐ Yes ☐ No ☐ DK
e. Sensorineural deafness on audiogram ☐ Yes ☐ No ☐ DK
f. Papilledema ☐ Yes ☐ No ☐ DK

17. Rheumatologic feature(s)
a. Arthralgia ☐ Yes ☐ No ☐ DK
b. Arthritis ☐ Yes ☐ No ☐ DK
c. If arthritis, specify joint(s): ☐ Hand ☐ Wrist ☐ Elbow ☐ Shoulder ☐ Spine ☐ Hip ☐ Knee ☐ Ankle ☐ Foot

18. Other features
a. History of premature delivery (< 37 weeks gestation) ☐ Yes ☐ No ☐ DK
b. Maternal history of pre-eclampsia ☐ Yes ☐ No ☐ DK
c. Periodic fevers ☐ Yes ☐ No ☐ DK
d. Frequency of periodic fevers (e.g. weekly, monthly):____

e. Height < 3rd percentile ☐ Yes ☐ No ☐ DK
f. Weight < 3rd percentile ☐ Yes ☐ No ☐ DK
g. Rash/CNS/joint features only present when exposed to cold temperatures ☐ Yes ☐ No ☐ DK
h. Family history of cryopyrin periodic fever syndrome ☐ Yes ☐ No ☐ DK
If YES specify relationship to patient (eg. mother, father, aunt etc.)____________________________

19. Investigations (ever present prior to onset of treatment)

a. Elevated neutrophil count (on at least 2 separate occasions when afebrile) ☐ Yes ☐ No ☐ Not Done ☐ DK
b. Elevated ESR (on at least 2 separate occasions when afebrile) ☐ Yes ☐ No ☐ Not Done ☐ DK
c. Elevated CRP (on at least 2 separate occasions when afebrile) ☐ Yes ☐ No ☐ Not Done ☐ DK
d. Bony overgrowth on plain X-ray of joint(s): ☐ Yes ☐ No ☐ Not Done ☐ DK
If yes which joint(s): ____

e. Other joint abnormalities on plain X-ray :____________________________________
f. Hydrocephalus on brain MRI/CT: ☐ Yes ☐ No ☐ Not Done ☐ DK
g. Other abnormalities on brain MRI/CT:____________________________________

20. CURRENT COMPLICATIONS/OUTCOMES

a. Concerns regarding developmental /cognitive delay ☐ Yes ☐ No ☐ DK
b. Requires hearing assisted device(s) (e.g.hearing aid, FM system) ☐ Yes ☐ No ☐ DK
c. Requires assistance with walking ☐ Yes ☐ No ☐ DK ☐ Not Applicable
d. Nephropathy/renal amyloidosis present ☐ Yes ☐ No ☐ DK
e. Requires/required intra-ventricular shunt due to hydrocephalus ☐ Yes ☐ No ☐ DK
f. Behavioural concerns (i.e. aggression, ADHD, depression) ☐ Yes ☐ No ☐ DK
If Yes Specify:____
g. Patient currently alive ☐ Yes ☐ No ☐ Lost to follow-up
h. If died, please specify age of death (years) ______and cause(s) of death:__________________________

21. CURRENT MEDICATIONS PRESCRIBED FOR TREATMENT OF CONDITION

Is the child currently on any medications for treatment of CAPS: ☐ Yes ☐ No ☐ DK *If YES which one(s)?*

Current medications	**Yes**	**No**	**DK**	**Approximate date or age commenced**
Corticosteroid	☐	☐	☐	☐☐/☐☐/☐☐ or __________ yrs of age
Anti-histamines	☐	☐	☐	☐☐/☐☐/☐☐ or __________ yrs of age
Anti-IL-1 receptor antagonist	☐	☐	☐	☐☐/☐☐/☐☐ or __________ yrs of age
Other (specify)__________________	☐	☐	☐	☐☐/☐☐/☐☐ or __________ yrs of age
Other (specify)__________________	☐	☐	☐	☐☐/☐☐/☐☐ or __________ yrs of age
Other (specify)__________________	☐	☐	☐	☐☐/☐☐/☐☐ or __________ yrs of age

22. IF ANTI-IL-1 RECEPTOR ANTAGONIST THERAPY BEING USED CURRENTLY

a. Name of anti-IL-1 receptor antagonist (e.g. Anakinra, Rilonacept, Canakinumab) :____________________
b. Current dose (mg/kg):_________; Route of administration: ☐ SC ☐ IV; Frequency of injections:________
c. Medications supply via: ☐ Drug company compassionate programme ☐ Hospital drug committee
d. Any complication(s) of the injections (e.g. pain):____________________________________

23. CURRENT CLINCIAL STATUS AT LAST EVALUATION

a. Date of last medical evaluation: ☐☐/☐☐/☐☐
b. Clinical/laboratory features at last evaluation:

Urticarial like rash:	☐ Yes	☐ No	☐ DK	Arthritis:	☐ Yes	☐ No	☐ DK
Arthralgia:	☐ Yes	☐ No	☐ DK	Chronic headache:	☐ Yes	☐ No	☐ DK
Deafness:	☐ Yes	☐ No	☐ DK	Macrocephaly:	☐ Yes	☐ No	☐ DK
Papilledema:	☐ Yes	☐ No	☐ DK	Hydrocephalus on CT/MRI:	☐ Yes	☐ No	☐ DK
Height < 3rd centile:	☐ Yes	☐ No	☐ DK	Weight < 3rd centile:	☐ Yes	☐ No	☐ DK
Periodic fevers:	☐ Yes	☐ No	☐ DK	Raised neutrophil count:	☐ Yes	☐ No	☐ DK
Raised ESR:	☐ Yes	☐ No	☐ DK	Raised CRP:	☐ Yes	☐ No	☐ DK

Thank you for completing this questionnaire. Please return it in the reply-paid envelope or fax to APSU 02 9845-3082.

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