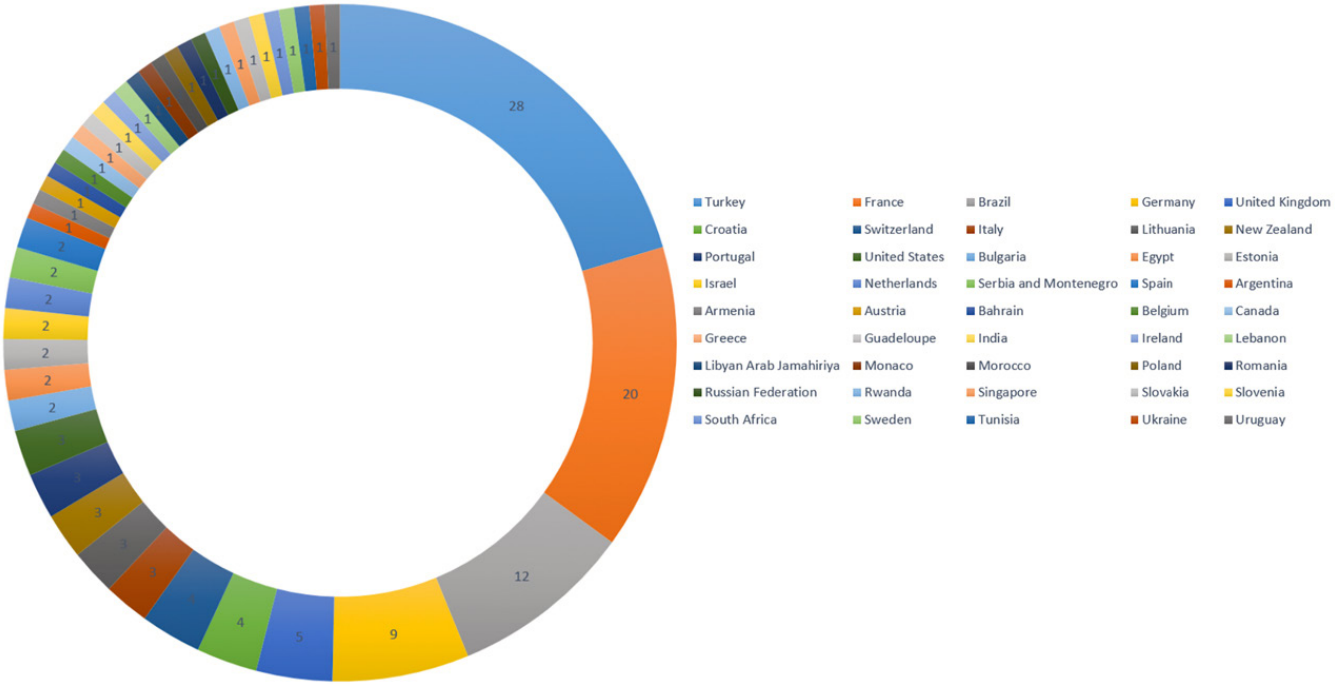

Supplementary Fig. S1. The number of participants per country (total number of respondents=137; total number of countries=45).

Supplementary Table S1. The survey questions that focus on demographic features and corticosteroid use in syndrome of undifferentiated recurrent fever (SURF).

What is your gender? (Female/Male/Do not wish to answer)
In which country is your institution?
What kind of institution do you work in?
- Private practice
- Hospital
- Tertiary hospital
- University hospital
- Other

Do you see outpatients, inpatients or both?
- Outpatients
- Inpatients
- Both

What kind of patients do you see?
- Only children
- Only adults
- Both children and adults

What is your specialty?
- Primary care general practitioner
- Primary care paediatrician
- Paediatric rheumatology
- Paediatric immunology
- Paediatric nephrology
- Paediatric cardiology
- Adult rheumatology
- Adult immunology
- Adult nephrology
- Adult cardiology
- Other

How many years have you cared for patients with recurrent fever?
- 0-4 years
- 5-10 years
- ≥10 years

Do you prescribe a treatment with a steroid dose at flare-onset? (Yes/No)

Please select the best treatment of steroids you will choose in SURF
- Prednisone/ Prednisolone/ Methylprednisolone/ Betamethasone/ Dexamethasone/ Deflazacort

---

How do you prescribe a treatment with a steroid dose at flare-onset in SURF?
- Routinely
- Not routinely
  - Depending on attack severity
  - Depending on family preferences
  - When antipyretics are not enough to control fever
  - Only for once, to confirm diagnosis
  - Depending on attack frequency
  - Other (please specify)

How many steroid doses per flare in SURF?
- Only 1 dose
- 1 or 2, depending on the response

Do you use any forms of steroids that are suitable for young children (*e.g.*, suspension, drops, etc.) in SURF? (Yes/No)

What is the maximum steroid uses that you recommend per year in SURF?
- <5
- 5-10
- >10

When do you consider that the patients responded to a steroid dose at flare-onset in SURF?
- Response within 3-4 hours
- Response within 12 hours
- Response within 24 hours

On which criteria would you consider that the patient did not respond in SURF?
- Need for more than 2 doses of steroid per episode
- No improvement in fever within 24 hours
- No improvement in fever within 12 hours
- Improvement in fever but fever recurs within the same episode after the steroid dose
- No improvement in fever within 3-4 hours
- Need for more than 1 dose of steroid per episode
- Other (please specify)

Steroids at flare-onset may decrease the intervals between the flares. If yes, based on which criteria would you consider another treatment in SURF?
- If decreased the quality of life
- Flare interval shorter than 2 weeks
- Decrease in attack intervals persisting for >3 months
- Flare interval shorter than 3 weeks
- Flare interval shorter than 4 weeks
- Decrease in attack intervals persisting for >6 months
- Only if severe attacks
- Decrease in attack intervals persisting for >1 year
- Other (please specify)

SURF: syndrome of undifferentiated recurrent fever.

Supplementary Table S2. The definitions of the syndrome of undifferentiated recurrent fever (SURF) or the inclusion criteria for SURF patients in the articles included during the literature review.

First author, year (ref. no.)	Definition of SURF or inclusion criteria for SURF patients
Harrison, 2016 (11)	1) Clinical features to suggest underlying SAIDs that could not be explained by autoimmune, infectious, or malignant processes; 2) Episodic or persistent constitutional symptoms (fever, malaise, and/or weight loss, or more organ-specific manifestations, such as sore throat, skin rashes, serositis, arthralgia, myalgia, lymphadenopathy, and other symptoms); 3) Absence of pathogenic mutations in known SAIDs genes and failure to meet the diagnostic criteria for defined polygenic autoinflammatory conditions
De Pauli, 2018 (8)	1) Clinical findings of hereditary periodic fever syndrome; 2) Absence of known causative mutation (negative results or polymorphisms of unknown significance)
Ter Haar, 2019 (15)	1) Fever episodes; 2) Elevated inflammatory markers; 3) Exclusion of other confounding conditions as well-defined SAIDs including PFAPA, infectious, autoimmune, neoplastic diseases; 4) Exclusion of patients with a positive genetic analysis that reveals a defined SAID or another diagnosis explaining their symptoms; 5) Exclusion of patients who met the clinical criteria for a defined monogenic autoinflammatory disease and not evaluated with further genetic tests
Papa, 2020 (5)	1) Symptoms related to SAIDs with a paediatric onset; 2) Exclusion of other common aetiologies; 3) Negative or not conclusive molecular diagnosis; 4) Exclusion of PFAPA syndrome
Luu, 2021 (13)	1) Recurrent fevers; 2) Normal growth; 3) Asymptomatic between episodes; 4) Absence of laboratory evidence of inflammation between episodes; 5) Absence of genetically defined SAIDs; 6) Absence of cyclic neutropenia, immunodeficiency, chronic infection, inflammatory bowel disease or autoimmunity
Kosukcu, 2021 (12)	1) Recurrent fever attacks; 2) High CRP levels along with clinical features of inflammation; 3) Absence of infections; 4) Absence of *MEFV* mutations; 5) Absence of known causative variants associated with monogenic SAIDs in WES
Marques, 2022 (14)	In this report, the authors presented four patients with SURF and gave their detailed clinical history. From the data they presented: • All patients had recurrent fever episodes and elevated acute phase reactants during these episodes. • Genetic test was negative in all four patients regarding monogenic SAIDs (in one patient the type of the genetic test was not specified; in three patients, SAID genetic screening panel was done with NGS.)
Gerritsma, 2023 (9)	1) Fever episodes; 2) Genetic analysis of at least two genes with no pathogenic or likely pathogenic variants; 3) Elevated inflammatory markers during episodes
Vitale, 2023 (16)	1) Febrile episodes; 2) Exclusion of patients with infections, autoimmune and neoplastic diseases; 3) Absence of genetic mutations related to defined SAIDs in NGS; 4) Exclusion of patients fulfilling any criteria for genetically determined FMF or any multifactorial autoinflammatory diseases, including PFAPA syndrome, Behçet's disease, Schnitzler syndrome, Castleman disease, or Still's disease
Gomez-Caverzaschi, 2024 (10)	1) Recurrent or persistent fever alone and/or other concomitant systemic or single-organ manifestations during at least 6 months; 2) Exclusion of alternative diagnoses of infectious, neoplastic, and autoimmune diseases; 3) Absence of pathogenic variants associated with monogenic SAIDs

CRP: C-reactive protein; FMF: familial Mediterranean fever; NGS: next generation sequencing; NI: not indicated; PFAPA: periodic fevers with aphthous stomatitis, pharyngitis, and adenitis; SAIDs: systemic autoinflammatory diseases; SURF: syndrome of undifferentiated recurrent fever; WES: whole exome sequencing.

Supplementary Table S3. The characteristics of patients with syndrome of undifferentiated recurrent fever (SURF) who received on demand corticosteroid treatment in the literature (except the total number of patients, all features delineated in the table belong to the SURF patients treated with on demand corticosteroids).

First author, year (ref. no.)	Total no. of patients	No. of patients treated with on demand CS	Adult or paediatric	Sex	Clinical features	Duration of febrile episode, days	Interval between febrile episodes, days	Type and dose of CS	Response to CS in a SURF episode	Concomitant therapy with CS
Harrison, 2016 (11)	11	1	Adult	F	Fever, abdominal pain, cervical lymphadenopathy, aphthous stomatitis, headache, malaise	3-5	28-42	Methylprednisolone; 1 mg/kg	No	Colchicine
De Pauli, 2018 (8)	23	21	Paediatric	NI	NC	NI	NI	Bethametasone; 0.1 mg/kg (n=11) 0.2-0.3 mg/kg (n=5), NI (n=5)	16/21 (76.2%)	Colchicine (n=13)
Ter Haar, 2019 (15)	187	104	Both	NC	NI	NI	NI	NI	85/104 (81.7%)	NI
Papa, 2020 (5)	34	18	Paediatric	NC	NI	NI	NI	NI	17/18 (94.4%)	NI
Luu, 2021 (13)	28	28	Paediatric	17M/11F	Fever (n=28), pharyngitis (n=8), cervical adenitis (n=7), aphthous stomatitis (n=5), abdominal pain (n=14), headache (n=12), ocular symptoms (n=6), nausea/vomiting (n=11), arthralgia (n=6), rash (n=3)	3.7-4.8 (2-7)*	42-67.6 (14-180)*	Prednisolone; ≤1 mg/kg	3/28 (10%)	NC
Kosukcu, 2021 (12)	7	1	Paediatric	1M	Fever, myalgia, abdominal pain, chest pain, arthralgia, fatigue	15-30	180	NI	Yes	None
Marques, 2022 (14)	4	2	Paediatric	2M	Fever (n=2), myalgia (n=2), abdominal pain (n=2), tonsillitis (n=1), aphthous stomatitis (n=2), pharyngitis (n=1), arthralgia (n=1), headaches (n=1), asthenia (n=1), erythema nodosum (n=1), maculopapular rash (n=1)	2, 3-5	30, 60	Prednisolone (n=2); 0.5 mg/kg (n=1)	1/2 (50%)	Colchicine (n=1)
Gerritsma, 2023 (9)	60	18/43	NI	NI	NI	NI	NI	NI	13/18	NI
Vitale, 2023 (16)	54	11	Adult	NC	NC	NC	NI	NI	NI	NC
Gomez-Caverzaschi, 2024 (10)	90	35	Adult	NI	NI	NI	NI	0.5-1 mg/kg	NI	NC

CS: corticosteroid; F: female; M: male; NC: not clear; NI: not indicated; SURF: syndrome of undifferentiated recurrent fever; *median (minimum-maximum); *mean range (minimum-maximum).