Suppl. Figure 1. The online survey on Management of urticarial vasculitis: A worldwide perspective

1 Your country of residence:
Please choose only one of the following:

Afghanistan	Comoros	Haiti
Algeria	Congo, Democratic	Honduras
American Samoa	Republic of the	Hong Kong
Andorra	Congo, Republic of	Hungary
Angola	the	Iceland
Antigua and Barbuda	Costa Rica	India
Argentina	Côte d'Ivoire	Indonesia
Armenia	Croatia	Iran
Australia	Cuba	Iraq
Austria	Cyprus	Ireland
Azerbaijan	Czech Republic	Israel
Bahamas	Denmark	Italy
Bahrain	Djibouti	Jamaica
Bangladesh	Dominica	Japan
Barbados	Dominican Republic	Jordan
Belarus	East Timor	Kazakhstan
Belgium	Ecuador	Kenya
Belize	Egypt	Kiribati
Benin	El Salvador	North Korea
Bermuda	Equatorial Guinea	South Korea
Bhutan	Eritrea	Kosovo
Bolivia	Estonia	Kuwait
Bosnia and	Ethiopia	Kyrgyzstan
Herzegovina	Faroe Islands	Laos
Botswana	Fiji	Latvia
Brazil	Finland	Lebanon
Brunei	France	Lesotho
Bulgaria	French Polynesia	Liberia
Burkina Faso	Gabon	Libya
Burundi	Gambia	Liechtenstein
Cambodia	Georgia	Lithuania
Cameroon	Germany	Luxembourg
Canada	Ghana	Macedonia
Cape Verde	Greece	Madagascar
Cayman Islands	Greenland	Malawi
Central African	Grenada	Malaysia
Republic	Guam	Maldives
Chad	Guatemala	Mali
Chile	Guinea	Malta
China	Guinea-Bissau	Marshall Islands
Colombia	Guyana	Mauritania

Mauritius
Mexico
Micronesia
Moldova
Monaco
Mongolia
Montenegro
Morocco
Mozambique
Myanmar
Namibia
Nauru
Nepal
Netherlands
New Zealand
Nicaragua
Niger
Nigeria
Northern Mariana
Islands
Norway
Oman
Pakistan
Palau
Palestine, State of
Panama
Papua New Guinea
Paraguay
Peru
Philippines
Poland
Portugal
Puerto Rico
Qatar
Romania
Russia
Rwanda
Saint Kitts and Nevis
Saint Lucia
Saint Vincent and the
Grenadines
Samoa
San Marino
Sao Tome and
Principe
Saudi Arabia
Senegal

Serbia
Seychelles
Sierra Leone
Singapore
Sint Maarten
Slovakia
Slovenia
Solomon Islands
Somalia
South Africa
Spain
Sri Lanka
Sudan
Sudan, South
Suriname
Swaziland
Sweden
Switzerland
Syria
Taiwan
Tajikistan
Tanzania
Thailand
Togo
Tonga
Trinidad and Tobago
Tunisia
Turkey
Turkmenistan
Tuvalu
Uganda
Ukraine
United Arab Emirates
United Kingdom
United States
Uruguay
Uzbekistan
Vanuatu
Vatican City
Venezuela
Vietnam
Virgin Islands, British
Virgin Islands, U.S.
Yemen
Zambia
Zimbabwe

2 Your specialty (Please choose all that apply):
- Allergy/Immunology
- Dermatology
- Paediatrics
- Rheumatology
- General practice
- Other (please specify): _____

3 How long have you been practicing in your specialty? (years):
_____

4 Your work environment (Please choose all that apply):
- Private practice
- University clinic
- Hospital
- Specialized urticaria centre
- Other (please specify): _____

5 How many Urticarial Vasculitis (UV) patients do you see per year? (Only numbers allowed): _____

6 Is the diagnosis of UV already established in most patients or do you diagnose these patients? Please choose only one of the following:
- The diagnosis of UV is already established in most patients
- I diagnose most patients

7 What are the reasons that patients with UV are referred to your institution? (Please choose all that apply):
- Establish a diagnosis
- Second opinion
- Treatment initiation
- Treatment optimization
- Clinical/basic research
- Other (please specify):_____

8 In how many of your UV patients do you observe the following <u>cutaneous</u> symptoms at any timepoint?

	No	≤20%	≤40%	≤60%	≤80%	<100%	All
Wheals	☐	☐	☐	☐	☐	☐	☐
Angioedema	☐	☐	☐	☐	☐	☐	☐
Itch	☐	☐	☐	☐	☐	☐	☐
Burning	☐	☐	☐	☐	☐	☐	☐
Localized pain	☐	☐	☐	☐	☐	☐	☐
Resolving of lesions with	☐	☐	☐	☐	☐	☐	☐

hyperpigmentation

	No	≤20%	≤40%	≤60%	≤80%	<100%	All
Other (please specify)_____	☐	☐	☐	☐	☐	☐	☐

9 In how many of your UV patients do you see the following <u>extracutaneous</u> symptoms at any timepoint?

	No	≤20%	≤40%	≤60%	≤80%	<100%	All
Constitutional symptoms (e.g. fatigue, asthenia, fever)	☐	☐	☐	☐	☐	☐	☐
Musculoskeletal involvement (e.g. arthralgia, arthritis)	☐	☐	☐	☐	☐	☐	☐
Ocular involvement (e.g. scleritis, uveitis)	☐	☐	☐	☐	☐	☐	☐
Lymph node enlargement	☐	☐	☐	☐	☐	☐	☐
Pulmonary involvement (e.g. COPD, dyspnea)	☐	☐	☐	☐	☐	☐	☐
GI involvement (e.g. pain, diarrhea, nausea)	☐	☐	☐	☐	☐	☐	☐
Kidney involvement (e.g. glomerular/tubulointerstitial nephritis)	☐	☐	☐	☐	☐	☐	☐
Ear, nose, and throat involvement (e.g. sinusitis, otitis media, hearing loss)	☐	☐	☐	☐	☐	☐	☐
Neurologic involvement	☐	☐	☐	☐	☐	☐	☐
Pericarditis	☐	☐	☐	☐	☐	☐	☐
Other (please specify)_____	☐	☐	☐	☐	☐	☐	☐

10 How do you diagnose UV? Please choose only <u>the three</u> most important signs and/or symptoms that apply:

- Wheals predominantly >24h
- Presence of post-inflammatory hyperpigmentation

- Systemic symptoms (e.g. fever, arthralgia, abdominal pain)
- Presence of underlying disease (e.g. malignancy, SLE)
- Histological analysis
- Low complement levels
- High levels of inflammation markers, e.g. ESR, CRP
- Poor response to antihistamines
- Other (please specify)_____

11 Which of the following tests do you order or perform in UV patients? Please choose all that apply:

	No	≤20%	≤40%	≤60%	≤80%	<100%	All patients
Skin biopsy	☐	☐	☐	☐	☐	☐	☐
Complete blood count	☐	☐	☐	☐	☐	☐	☐
Erythrocyte sedimentation rate	☐	☐	☐	☐	☐	☐	☐
C-reactive protein	☐	☐	☐	☐	☐	☐	☐
Anti-thyroid antibodies	☐	☐	☐	☐	☐	☐	☐
Complement component (C3, C4, C1q, CH50)	☐	☐	☐	☐	☐	☐	☐
Thyroid stimulating hormone	☐	☐	☐	☐	☐	☐	☐
Eosinophil cationic protein	☐	☐	☐	☐	☐	☐	☐
D-dimer	☐	☐	☐	☐	☐	☐	☐
Serological testing for hepatitis	☐	☐	☐	☐	☐	☐	☐
Search for malignancies	☐	☐	☐	☐	☐	☐	☐
Search for allergy (skin tests, allergen-specific IgE, total IgE)	☐	☐	☐	☐	☐	☐	☐
Antinuclear antibodies	☐	☐	☐	☐	☐	☐	☐
Autologous serum skin test	☐	☐	☐	☐	☐	☐	☐
Specific autoantibodies*	☐	☐	☐	☐	☐	☐	☐
None	☐	☐	☐	☐	☐	☐	☐

Other (please specify)_____ ☐ ☐ ☐ ☐ ☐ ☐ ☐

*e.g. anti-SSA/B, anti-dsDNA, anti-RNP, anti-SM, anti-C1q, ANCAs

12 What comorbidities/potential causes of UV do you see in your patients?

	None	≤20%	≤40%	≤60%	≤80%	<100%	Always
Idiopathic UV	☐	☐	☐	☐	☐	☐	☐
Malignancy	☐	☐	☐	☐	☐	☐	☐
Systemic lupus erythematosus	☐	☐	☐	☐	☐	☐	☐
Other autoimmune diseases	☐	☐	☐	☐	☐	☐	☐
Hepatitis C	☐	☐	☐	☐	☐	☐	☐
Other chronic infections	☐	☐	☐	☐	☐	☐	☐
Drugs	☐	☐	☐	☐	☐	☐	☐
I don't usually look for underlying causes of UV	☐	☐	☐	☐	☐	☐	☐
Other (please specify)_____	☐	☐	☐	☐	☐	☐	☐

13 Which of the following medications do you use in UV? Please choose all that apply:

	1st line	2nd line	3rd line	≥4th line	Do not use
First-generation H1-antihistamines	☐	☐	☐	☐	☐
Second-generation H1-antihistamines at standard dose	☐	☐	☐	☐	☐
Updosed second-generation H1-antihistamines (2-4 times the standard dose)	☐	☐	☐	☐	☐
H2-antihistamines (e.g. famotidine or ranitidine)	☐	☐	☐	☐	☐

Cyclosporine	☐	☐	☐	☐	☐
Omalizumab	☐	☐	☐	☐	☐
Montelukast	☐	☐	☐	☐	☐
NSAIDs	☐	☐	☐	☐	☐
Glucocorticosteroids	☐	☐	☐	☐	☐
Azathioprine	☐	☐	☐	☐	☐
Methotrexate	☐	☐	☐	☐	☐
Rituximab	☐	☐	☐	☐	☐
Anakinra	☐	☐	☐	☐	☐
Canakinumab	☐	☐	☐	☐	☐
Mycophenolate mofetil	☐	☐	☐	☐	☐
Intravenous immunoglobulin	☐	☐	☐	☐	☐
Dapsone	☐	☐	☐	☐	☐
Colchicine	☐	☐	☐	☐	☐
Hydroxychloroquine	☐	☐	☐	☐	☐
Pentoxifylline	☐	☐	☐	☐	☐
Plasmapheresis	☐	☐	☐	☐	☐
Other (please specify)_____	☐	☐	☐	☐	☐

14 What is(are) your greatest challenge(s) in managing UV patients? (please choose all that apply)
- No clinical guidelines and treatment algorithms exist
- Clinical diagnostic criteria are not clear
- Histological diagnostic criteria are not clear
- Many drugs have limited efficacy
- Many drugs have potentially serious adverse effects
- Often severe and difficult-to-treat disease
- Need for help from other specialists, especially in the case of underlying disease
- Novel treatment is not available or costs too high in my country of residence
- I don't have enough clinical experience in the management of UV
- It is difficult to find an underlying diseases (a cause of UV is usually unknown)
- Other (please specify): _____

7

Thank you for taking your time to participate in our survey! You helped to change the future of the management of urticarial vasculitis patients!

If you have comments or suggestions for us, please provide them here:

Thank you very much for your participation!