SARCOIDOSIS VASCULITIS AND DIFFUSE LUNG DISEASES 2014; 31; 19-27

The WASOG Sarcoidosis Organ Assessment Instrument: An update of a previous clinical tool

*M.A. Judson[1], U. Costabel[2], M. Drent[3], A. Wells[4], L. Maier[5], L. Koth[6], H. Shigemitsu[7], D.A. Culver[8], J.Gelfand[6], D. Valeyre[9], N. Sweiss[10], E. Crouser[11], A. S. Morgenthau[12], E.E. Lower[13], A. Azuma[14], M. Ishihara[15], S. Morimoto[16], T. Yamaguchi[17], N. Shijubo[18], J.C. Grutters[19], M. Rosenbach[20], H.P. Li[21], P. Rottoli[22], Y. Inoue[23], A. Prasse[24], R.P. Baughman[13], The WASOG Sarcoidosis Organ Assessment Instrument Investigators**

[1]Albany Medical College; Albany, New York USA; [2]Ruhrlandklinik, University Hospital, University Duisburg-Essen, Essen, Germany; [3]Gelderse Vallei Hospital Ede; Maastricht University, Maastricht, The Netherlands; [4]Royal Brompton Hospital, London; [5]National Jewish Health, Denver, Colorado USA; [6]University of California, San Francisco, San Francisco, CA USA; [7]University of Southern California Keck School of Medicine, Los Angeles, California USA; [8]Cleveland Clinic, Cleveland, Ohio USA; [9]AP-HP, Hôpital Avicenne, Université Paris 13, Sorbonne Paris Cité, France; [10]University of Illinois College of Medicine at Chicago, Chicago, Illinois USA; [11]The Ohio State University Medical Center, Columbus, Ohio USA; [12]Icahn School of Medicine at Mount Sinai; New York, New York USA; [13]University of Cincinnati Medical Center; Cincinnati, OH, USA; [14]Nippon Medical School, Tokyo, Japan; [15]Department of Ophthalmology and Visual Science, Yokohama City, Japan; [16]Aoyama General Hospital, Tokyo, Japan; [17]Japan Railway Tokyo General Hospital, Tokyo, Japan; [18]JR Sapporo Hospital, Sapporo, Japan; [19]St. Antonius Hospital, Utrecht, The Netherlands; [20]University of Pennsylvania School of Medicine, Philadelphia, Pennsylvania USA; [21]Tongji University, School of Medicine, Shanghi, China; [22]Azienda Ospedaliera Universitaria Senese, Italy; [23]Kinki-Chuo Chest Medical Center, Osaka, Japan; [24]University Hospital, Freiburg, Germany; * see appendix 1

Abstract. *Introduction:* A Case Control Etiology of Sarcoidosis Study (ACCESS) sarcoidosis organ assessment instrument has been used for more than a decade to establish uniform standards for the probability of sarcoidosis organ involvement. The ACCESS instrument has become increasingly outdated as new technologies have been developed. Furthermore, the ACCESS instrument failed to address all possible organs involved with sarcoidosis. For these reasons, the World Association of Sarcoidosis and Other Granulomatous Diseases (WASOG) developed a new sarcoidosis organ assessment instrument. *Methods:* Clinical sarcoidosis experts assessed various clinical manifestations for the probability of sarcoidosis organ involvement. Two criteria were required to apply this assessment: 1) histologic evidence of granulomatous inflammation of unknown cause in an organ that was not being assessed; 2) the clinical manifestation being addressed required that alternative causes other than sarcoidosis had been reasonably excluded. Clinical manifestations were assessed as either: a) highly probable: likelihood of sarcoidosis causing this manifestation of at least 90%.; b) probable: likelihood of sarcoidosis causing this manifestation of between 50 and 90%; c) possible: likelihood of sarcoidosis causing this manifestation of less than 50%. The sarcoidosis experts voted on the likelihood of sarcoidosis causing each manifestation using Delphi study methodology where at least 70% agreement of the experts was needed for consensus. *Results:* Various clinical manifestations were classified as highly probable, at least probable, possible, or indeterminate when no consensus could be reached. *Conclusion:* An instrument was developed by expert opinion that may be useful for the clinician and researcher in establishing criteria for sarcoidosis organ involvement.

(Sarcoidosis Vasc Diffuse Lung Dis 2014; 31: 19-27)

Received: 01 November 2013
Accepted after revision: 9 December 2013
Correspondence: Marc A. Judson, M.D.
Division of Pulmonary and Critical Care Medicine MC-91, Albany Medical College, 47 New Scotland Avenue, Albany, New York 12208 USA.
E-mail: judsonm@mail.amc.edu

Key words: sarcoidosis, organ, diagnosis, consensus

Introduction

Sarcoidosis is a multisystem granulomatous disease of unknown cause.[1] Granulomatous inflammation from sarcoidosis may occur in any organ. ACCESS (A Case Control Etiology of Sarcoidosis Study) was a study of sarcoidosis in the 1990's funded by the National Institutes of Health that was primarily aimed at searching for the etiology of sarcoidosis. For the purposes of that effort, the ACCESS investigators were concerned with establishing criteria for sarcoidosis organ involvement. They had noticed that a large number of sarcoidosis clinical trials did not describe rigorous entry criteria for enrollment; other trials that did clearly describe entry criteria for various sarcoidosis organ involvements were not consistent with each other. This issue was the major impetus for the development of the ACCESS Sarcoidosis Organ Assessment Instrument.[2] The instrument assessed various clinical findings of 15 organs in terms of their likelihood of representing sarcoidosis. These clinical findings were graded as "definite," "probable," and "possible" evidence of organ involvement with sarcoidosis. For the purposes of the ACCESS study, "definite" and "probable" involvement was considered to represent organ involvement with sarcoidosis. A prerequisite for using the ACCESS instrument to evaluate the likelihood of a clinical finding of an organ representing sarcoidosis was that at least one additional organ had demonstrated granulomatous inflammation of no alternative cause.

The ACCESS instrument, although useful, suffers from some deficiencies. First, it was developed more than one decade ago, and it is somewhat outdated as new technologies have been developed in the interim for the diagnosis and monitoring of sarcoidosis. Second, the instrument failed to cover all possible organs involved with sarcoidosis. Third, several common and very specific manifestations of sarcoidosis were not addressed. For these reasons, the World Association of Sarcoidosis and Other Granulomatous Disorders (WASOG) developed a new sarcoidosis organ assessment instrument. This manuscript will describe this instrument.

Methods

Clinical sarcoidosis experts who were members of WASOG were invited to participate in development of the instrument (Appendix 1). Individuals were invited to serve as "organ group leaders," who would be responsible for developing criteria for a specific organ. Other WASOG members were invited to participate as members of any organ group that was of interest to them. In addition, the organ group leaders were given the authority to invite non-WASOG members to their groups if they believed such individuals had clinical expertise in the assessment of sarcoidosis involvement of specific organs. A list of the organ group leaders and members is shown in Appendix 1.

Various clinical manifestations were assessed for the probability of organ involvement. Two criteria were required to be fulfilled in order to apply this assessment: 1) histologic evidence of granulomatous inflammation of unknown cause needed to be demonstrated in at least one organ that was not being assessed; 2) the clinical manifestation being assessed required that all alternative causes other than sarcoidosis for this clinical manifestation had been reasonably excluded. Provided that these two criteria were fulfilled, clinical manifestations were assessed by assigning them to one of three categories, highly probable, probable, or possible, interpreted as follows. **Highly probable**: such a manifestation is highly specific for sarcoidosis, with a likelihood of sarcoidosis causing this manifestation of at least 90%. In such cases, organ involvement may be assumed without a biopsy. **Probable**: such a manifestation is fairly specific for sarcoidosis, with a likelihood of sarcoidosis causing this manifestation of between 50 and 89%. In general, organ involvement in this category would be adequate to establish a clinical diagnosis of sarcoidosis in that organ. **Possible**: such a manifestation is not specific for sarcoidosis. For all organs, a biopsy showing granulomatous inflammation where alternative causes were reliably excluded was considered "highly probable"; therefore, such a biopsy was not included in this instrument.

Each organ group developed a list of common clinical conditions in their specific organ that could be considered as representing organ involvement with sarcoidosis. The probability of each of these clinical conditions as representing sarcoidosis was determined through organ group discussion that occurred via email and/or conference calls. Although organ group participants were encouraged to use the limited medical evidence available, these criteria were established by expert opinion. Other groups relied on voting of the members. After all organ assessments were developed

by each organ group, each of the clinical conditions was presented to all the organ group leaders and other sarcoidosis experts (all the authors of this manuscript) for a vote as to whether that clinical condition represented "highly probable," "probable," or "possible" involvement of sarcoidosis in that organ (vide supra). The voting group was blinded as to the assessments made by each specific organ group. It was this final vote by which the determination of likelihood of organ involvement on the basis of each clinical condition was based.

In terms of the voting, Delphi study methodology was used to determine that consensus was reached in that at least 70% of the experts needed to agree for consensus.[3] If at least 70% of the experts voted that the clinical condition was "highly probable" to represent sarcoidosis, a consensus was reached. If fewer than 70% of the experts voted that the clinical condition was "highly probable" but at least 70% of the experts voted that it was "highly probable" or "probable," then a consensus was reached that the clinical condition was at least probable. If at least 70% of the experts voted that the clinical condition was only "possible" to represent sarcoidosis, a consensus was reached. Finally, if less than 70% of the experts agreed that the clinical condition was a) "highly probable," b)"highly probable" or "probable" or c)"possible, " then a consensus was not reached. In this case, it was unclear, in the opinion of these experts, if such a clinical condition was adequate or inadequate to represent organ involvement with sarcoidosis. After all these expert opinions were rendered, this manuscript was written and was presented to the Executive Committee of WASOG for editing, comments, and approval.

The following organs were evaluated in this instrument: lung, skin, eye, liver, calcium dysregulation, neural tissue, kidney, heart, peripheral lymph node, bone marrow, spleen, bone/joint, ear-nose-throat, parotid/salivary glands, and muscle. In addition, a category of "other organs" was created to encompass all organs involved other than the 15 specific ones listed above.

Results

Table 1 shows the characteristic of the experts. All but one cared for more than 50 sarcoidosis patients yearly. Almost all had participated in at least one clinical sarcoidosis trial and more than three-quarters had published more than 10 manuscripts concerning sarcoidosis.

Table 2 shows the voting results and consensuses reached in terms of the likelihood of various clinical manifestations representing specific organ involvement with sarcoidosis. For all of the 16 organs, the experts reached consensus that at least one clinical condition was as "at least 'probable'" as representing sarcoidosis. Although several of these clinical conditions concerned laboratory testing, several of them concerned physical examination findings or patient symptoms.

Discussion

We have proposed the WASOG Organ Assessment Instrument as an update of the original ACCESS organ assessment instrument[2] based on improvement in diagnostic testing for sarcoidosis and new medical evidence that has occurred over the previous decade. Although this document is based as much as possible on medical evidence, it also incorporates expert opinion of WASOG members and

Table 1. Characteristics of the Experts

Number of years since training completed (N, %):	
<5	2, 8%
5-10	3, 12%
10-20	6, 23%
>20	15, 58%
Number of sarcoidosis patients treated per year, on average:	
0 – 50	1, 4%
51 – 100	6, 23%
101 – 250	11, 42%
>250	8, 31%
Number of sarcoidosis clinical trials that you have participated in:	
0	3, 12%
1-3	9, 35%
4-10	10, 38%
>10	4, 15%
Number of publications authored or co-authored concerning sarcoidosis:	
0	0, 0%
1-10	6, 23%
11-20	4, 15%
>20	16, 62%

Table 2. Histological types of cancer in the "sarcoidosis and cancer" group.

	Highly Probable	At Least Probable	Possible	No Consensus
Lung	CXR: bilateral hilar adenopathy (19-2-0) Chest CT: perilymphatic nodules (18-2-1) Chest CT: symmetrical hilar/mediastinal adenopthy (21-0-0) PET/Gallium-67: mediastinal/hilar enhancement (17-4-0)	CXR: diffuse infiltrates (4-13-3) CXR: upper lobe fibrosis (9-10-2) Chest CT: peribronchial thickening (10-8-3) BAL: lymphocytic alveolitis (6-14-1) BAL: elevated CD4/CD8 ratio (11-9-1) PET/Gallium-67: diffuse parenchymal lung enhancement (9-8-4) TBNA: lymphoid aggregates/giant cells (7-8-5)	CXR: localized infiltrate (1-2-18) PFT: obstruction (1-2-17)	PFT: restriction (2-6-12) PFT: isolated reduction in diffusing capacity (2-6-12)
Skin	Lupus pernio (16-2-0)	Subcutaneous nodules or plaques (3-14-1) Inflammatory papules within a scar or tattoo (7-8-2) Violaceous or erythematous annular lesions (2-15-3) Violaceous or erythematous macular, papular lesions around the eyes, nose, or mouth (11-3-3)	Atypical lesions: ulcerative, erythrodermic, alopecic, ichthyosiform (0-2-14)	Verrucous/scaly papules or plaques (3-7-7) Hypo- or Hyperpigmented macules or patches (2-8-7)
Liver		Abdominal imaging demonstrating hepatomegaly (1-12-5) Abdominal imaging demonstrating hepatic nodules (3-13-1)		Hepatomegaly on physical examination (0-10-8) Serum alkaline Phosphate > 3X the upper limit of normal (3-8-6)
Eye	uveitis (16-1-0) optic neuritis (13-2-2) mutton fat keratic precipitates (12-1-3) iris nodules (13-3-0) snowball/string of pearls (pars planitis) (10-3-1)	lacrimal gland swelling (10-4-3) trabecular meshwork nodules (9-6-0) retinitis (5-9-0) scleritis (5-7-2) multiple chorioretinal peripheral lesions (6-8-1) adnexal nodularity (8-5-2) candle wax drippings (11-2-3)	cataract (0-1-15) glaucoma (1-3-12) red eye (0-3-14)	blindness (0-7-10) painful eye (0-5-10) cystoid macular edema (2-7-5)
Spleen		Low attenuation nodules on CT (7-11-1) PET/gallium-67 uptake in splenic nodules (7-11-1) Splenomegaly on imaging or physical examination (4-11-4)		
Salivary Gland	Positive gallium-67 scan ("Panda sign") (14-3-0) Positive PET scan of the parotid glands (12-5-0)	Symmetrical parotitis with syndrome of mumps (10-5-2) Enlarged salivary glands (2-10-5)	Dry mouth (2-3-12)	

Continued

Table 2. (Continued)

ENT		Granulomatous changes on direct laryngoscopy (12-5-1) Consistent imaging studies (e.g. sinonasal erosion, mucoperiosteal thickening, positive PET scan) (6-8-4)	chronic sinusitis (0-1-17)	Nasal crusting, epistaxis, or anosmia associated with chronic sinus congestion (1-9-8)
Calcium-VitD	hypercalcemia plus all of the following: a) a normal serum PTH level; b) a normal or increased 1,25-OH dyhydroxy vitamin D level; c) a low 25-OH vitamin D level (17-2-0) hypercalciuria plus all of the following: a) a normal serum PTH level; b) a normal or increased 1,25-OH dyhydroxy vitamin D level; c) a low 25-OH vitamin D level (16-3-0)	nephrolithiasis plus all of the following: a) a normal serum PTH level; b) a normal or increased 1,25-diOH vitamin D level; c) a low 25-OH vitamin D level (12-7-0) hypercalciuria without serum PTH and 25 and 1,25 vitamin D levels (3-11-5) nephrolithiasis with calcium stones, without serum PTH and 25 and 1,25 vitamin D levels (4-11-4)	Nephrolithiasis, no stone anlaysis (1-3-15)	
Bone-Joint	Typical radiographic features (trabecular pattern, osteolysis, cysts/punched out lesions) (16-3-0)	Dactylitis (10-6-2) Nodular tenosynovitis (4-9-4) Positive PET, MRI, or gallium-67 bone imaging (8-9-1)	Arthralgias (0-5-14)	Non-specific arthritis (1-6-12)
Bone Marrow	PET displaying diffuse uptake (13-4-2)			leukopenia (2-8-8) anemia (1-5-13) thrombocytopenia (1-5-13)
Muscle		Positive imaging (MRI, Gallium-67) (13-7-0) Palpable muscle masses (3-11-6)	Myalgias (0-6-14)	Elevated serum muscle enzymes (5-8-7)
Extra-Thoracic Lymph Node		Multiple enlarged palpable cervical or epitrochelar lymph nodes without B symptoms (5-13-1) Enlarged lymph nodes identified by imaging in at least 2 peripheral or visceral lymph node stations without B symptoms (5-14-1)		Multiple enlarged palpable peripheral or visceral lymph nodes with B symptoms (1-10-9) Multiple palpable enlarged peripheral or visceral lymph nodes at sites other than cervical and epitrochlear (2-10-7)
Kidney		Treatment-responsive renal failure with no other risk factors. (9-9-1) Treatment-responsive renal failure in patient with diabetes and/or hypertension. (0-12-5)	Renal failure with other potential risk factors (0-4-15)	CT evidence of abnormal renal enhancement. (0-12-7)

Continued

Table 2. (Continued)

Nervous System	Clinical syndrome consistent with granulomatous inflammation of the meninges, brain, ventricular (CSF) system, cranial nerves, pituitary gland, spinal cord, cerebral vasculature or nerve roots -plus- An abnormal MRI characteristic of neurosarcoidosis, defined as exhibiting abnormal enhancement following the administration of gadolinium or a cerebrospinal fluid exam demonstrating inflammation (17-3-0)	Isolated facial palsy, negative MRI (6-8-5) Clinical syndrome consistent with granulomatous inflammation of the meninges, brain, ventricular (CSF) system, cranial nerves, pituitary gland, spinal cord, cerebral vasculature, nerve roots but without characteristic MRI or CSF findings (4-11-4)	Seizures, negative MRI (0-3-15) Cognitive decline, negative MRI (0-1-17)	Peripheral neuropathy involving large fibers (including axonal and demyelinating polyneuropathies and multiple mononeuropathies) (4-9-6) Cranial nerve palsies other than VII, negative MRI (4-7-8) Pleocytosis in the CSF (1-7-10) Low CSF glucose (0-6-12)
Cardiac		Treatment responsive CM or AVNB (12-7-1) Reduced LVEF in the absence of other clinical risk factors (2-13-4) Spontaneous or inducible sustained VT with no other risk factor (6-12-1) Mobitz type II or 3rd degree heart block (11-6-2) Patchy uptake on dedicated cardiac PET (10-8-1) Delayed enhancement on CMR (12-5-1) Positive gallium uptake (8-11-0) Defect on perfusion scintigraphy or SPECT scan (4-11-3) T2 prolongation on CMR (2-11-5)	Reduced LVEF in the presence of other risk factors (e.g., HTN, DM) (0-1-17) Atrial dysrhythmias (0-4-15)	Frequent ectopy (>5% QRS) (0-6-13) Bundle branch block (2-8-9) Impaired RV function with a normal PVR (0-8-10) Fragmented QRS or pathologic Q waves in ≥2 anatomically contiguous leads (0-7-10) At least one abnormal SAECG domain (0-6-10) Interstitial fibrosis or monocyte infiltration (4-8-7)
Other Organs		Positive imaging (3-8-3)		

**: at least 70% agreement by the experts*
***: for all clinical conditions, a) biopsy of that organ demonstrating granulomatous inflammation of no alternate cause implies highly probable involvement, b) another organ has demonstrated granulomatous inflammation of no alternate cause, c) alternative causes for the clinical manifestation have been reasonable excluded; CXR: chest radiograph; PFT: pulmonary function tests; Chest CT: chest computed tomography scan; TBNA: transbronchial needle aspiration (of a mediastinal lymph node); PET: positron emission tomography scan; Gallium-67: Gallium-67 nuclear scan; BAL: bronchoalveolar lavage; 3X: three times; PTH: serum parathyroid hormone; ENT: ear, nose, throat; Vit D: vitamin D; OH: hydroxy; di-OH: di-hydroxy; MRI: magnetic resonance imaging; B symptoms: fever, weight loss, or night sweats; CSF: cerebral spinal fluid; CM: cardiomyopathy; AVNB: atrioventricular nodal block; LVEF: left ventricular ejection fraction; HTN: systemic hypertension; DM: diabetes mellitus; VT: ventricular tachycardia; RV: right ventricular; SAECG: signal-averaged electrocardiogram; CMR: cardiovascular magnetic resonance imaging*

others who have expertise in the various organ manifestations of sarcoidosis. Unlike the original ACCESS organ assessment instrument, an organized process including a blinded vote was used to reach a consensus of the experts. In addition, the category of "definite" organ involvement in the ACCESS instrument was changed to "highly probable" because even histologic evidence of granulomatous inflammation is not definitive for the diagnosis of sarcoidosis.

This instrument should be viewed as a tool to assign probability to specific clinical findings as representing organ involvement with sarcoidosis. Many believe that because sarcoidosis is a multisystem disease, the diagnosis requires the presence of granulomatous inflammation in at least two organs.[1,4] It is unclear if this requirement is universally agreed upon. Regardless, this instrument assigns a probability for an additional organ having sarcoidosis based on clin-

ical criteria if another organ has demonstrated granulomatous inflammation of unknown cause previously. For clinicians who require that two organs demonstrate granulomatous inflammation of unknown cause for sarcoidosis to be diagnosed, this instrument would allow the diagnosis of sarcoidosis to be established in many cases without the need to biopsy a second organ.

This instrument is not designed to be used to assess activity or severity of sarcoidosis. Furthermore, this instrument is not a suggested algorithm to detect specific sarcoidosis organ involvement. In most cases, sarcoidosis organ involvement that does not cause significant symptoms does not require therapy.[5] Therefore, there is little reason in most cases to pursue a diagnosis of sarcoidosis in every possible organ that may be involved with the disease. Organ involvement may be occult without causing any clinical manifestations, and we are not advocating using this instrument to determine if organ involvement is clinically significant. In addition, the instrument is not designed to determine the need for treatment. It may be appropriate to treat clinical findings meeting only possible involvement criteria if the clinician determines that this is warranted.

This instrument may give guidance as to whether a clinical diagnosis of sarcoidosis organ involvement can be made without performing a biopsy to demonstrate granulomatous inflammation. Taking these individual clinical scenarios in isolation without regard to other clinical findings, we would propose that highly probable or at least probable involvement suggests that scenario is adequate for a clinical diagnosis of organ involvement. We acknowledge that the presence of a scenario voted as possible involvement may be adequate for a clinical diagnosis of sarcoidosis if additional other clinical findings are present.

There are several limitations of this instrument. First, the likelihood of each clinical finding described in the instrument is assigned a probability of representing sarcoidosis involvement of an organ based on the assumption that all other alternative causes for that clinical finding have been "reasonably excluded." This instrument provides no metric for this process, so that the method of excluding alternative diagnoses is arbitrary. At a minimum, attempts should be made to exclude mycobacterial infection, fungal infection, and malignancy. We acknowledge that if a very rigorous process is made to exclude alternative causes for the clinical findings discussed, that the likelihood of sarcoidosis could potentially be "upgraded." Second, there is no evidence that this instrument identifies sarcoidosis phenotypes that relate to specific genotypes or other specific mechanisms of disease. Other instruments have demonstrated evidence of such associations, albeit weakly.[6] It is possible that this instrument might function similarly, but that remains conjectural at this time. Third, the organ manifestations of sarcoidosis in our instrument are not comprehensive. Therefore, several manifestations were not appraised by the experts and, therefore, are unclassified. In addition, this instrument did not evaluate "para-sarcoidosis syndromes" that are often of major concern to sarcoidosis patients. These are conditions found frequently in sarcoidosis patients but are not directly attributable to granulomatous organ involvement and include small fiber neuropathy,[7-9] fatigue,[10-13] depression[13-15] and constitutional symptoms such as fever, weight loss, and malaise.[16] Finally, similar to our comments concerning the need for a biopsy in the preceding paragraph, each of the clinical manifestations that we assessed in this instrument does not always occur in isolation. It is possible that if a patient has evidence of multiple manifestations, each of which we regard as "probable" sarcoidosis, this may raise the probability of sarcoidosis to "highly probable." However, such an analysis is too complex to be examined presently.

We acknowledge that our position that highly probable or probable organ involvement is adequate for a clinical diagnosis of sarcoidosis involvement in an organ is arbitrary. Some may prefer to be more rigorous and require that organ involvement be highly probable for sarcoidosis organ involvement to be assumed without performing a confirmatory biopsy. For these reasons, Table 1 supplies the votes of all the experts for each clinical condition and designates the clinical conditions where a consensus was reached that they were highly probable.

In summary, we have presented an instrument that we consider useful in assessing the probability of organ involvement with sarcoidosis. Although we believe that this instrument will be useful for the clinician and clinical researcher involved with sarcoidosis patients, we suspect that it will require further modification over time as additional diagnostic tests are developed and new medical evidence is generated.

Appendix 1: Organ Groups for initial establishment of clinical scenarios for future voting

Lung
Lead: Robert Baughman. Members: Norman Soskel, Athol Wells, Elliott Crouser, Laura Koth, Marjolein Drent, Paola Rittoli, Daniel Culver, Milton Rossman, Ulrich Costabel, Lisa Maier, Dominique Valeyre, Hide Shigemitsu, Nadera Sweiss, Dominique Israel-Biet, Manuel Riberto Neto, Dheeraj Gupta, Eva Carmona; Patterson, Karen, Andrew P. Matragrano

Skin
Lead: Misha Rosenbach. Members: Marc Judson, Gloria Westney, Debasis Sahoo

Eye
Lead: Robert Baughman. Members: Elyse Lower, Adam Morgenthau

Liver
Lead: Adam Morgenthau. Members: Marjolein Drent, Gloria Westney, Lisa Maier; Nadera Sweiss

Calcium
Lead: Marc Judson. Members: Lisa Maier, Laura Koth, Hide Shigemitsu; Nadera Sweiss

Neuro
Lead: Jeffery Gelfand. Members: Marjolein Drent, Barney Stern, Jinny Tavee, Elske Hoitsma, Hide Shigemitsu, Kenkichi Nozaki, Fleur Cohen Aubart

Kidney
Lead: Elliott Crouser. Members: Milton Rossman, Daniel Culver; Nadera Sweiss

Heart
Lead: Daniel Culver. Members: Elliott Crouser, Nabeel Hamzeh, Milton Rossman, Ulrich Costabel, Vasanth Vedantham, Lisa Maier, Adam Morgenthau, Catherine Chapelon, David Bernie, Debabrata Bandyopadhyay

Peripheral Lymph Node
Lead: Lower. Member: Marc Judson

Bone Marrow
Lead: Adam Morgenthau. Members: Elyse Lower

Spleen
Lead: Elyse Lower. Members: Gloria Westney, Adam Morgenthau

Bone/Joint
Lead: Nadera Sweiss. Members: Laura Koth, Debasis Sahoo, Andrew Gross ; Arthur Yee

ENT
Lead: Marc Judson. Members: Gloria Westney, Lisa Maier

Parotid/Salivary Glands
Lead: Robert Baughman. Member: Debasis Sahoo

Muscle
Lead: Dominique Valeyre. Members: Marjolein Drent, Nadera Sweiss, Arthur Yee

Other organs
Lead: Marc Judson. Member: Robert Baughman

References

1. Hunninghake GW, Costabel U, Ando M, et al. ATS/ERS/WASOG statement on sarcoidosis. American Thoracic Society/European Respiratory Society/World Association of Sarcoidosis and other Granulomatous Disorders. Sarcoidosis Vasc Diffuse Lung Dis 1999; 16:149-173
2. Judson MA, Baughman RP, Teirstein AS, et al. Defining organ involvement in sarcoidosis: the ACCESS proposed instrument. ACCESS Research Group. A Case Control Etiologic Study of Sarcoidosis. Sarcoidosis Vasc Diffuse Lung Dis 1999; 16:75-86
3. de Meyrick J. The Delphi method and health research. Health Educ 2003; 103:7-16
4. Judson MA. The diagnosis of sarcoidosis. Clin Chest Med 2008; 29:415-427, viii
5. Baughman RP, Culver DA, Judson MA. A concise review of pulmonary sarcoidosis. Am J Respir Crit Care Med 2011; 183:573-581
6. Prasse A, Katic C, Germann M, et al. Phenotyping sarcoidosis from a pulmonary perspective. Am J Respir Crit Care Med 2008; 177:330-336

7. Hoitsma E, Drent M, Verstraete E, et al. Abnormal warm and cold sensation thresholds suggestive of small-fibre neuropathy in sarcoidosis. Clin Neurophysiol 2003; 114:2326-2333
8. Bakkers M, Merkies IS, Lauria G, et al. Intraepidermal nerve fiber density and its application in sarcoidosis. Neurology 2009; 73:1142-1148
9. Judson MA. Small fiber neuropathy in sarcoidosis: Something beneath the surface. Respir Med 2011; 105:1-2
10. Drent M, Lower EE, De Vries J. Sarcoidosis-associated fatigue. Eur Respir J 2012; 40:255-263
11. de Kleijn WP, De Vries J, Lower EE, et al. Fatigue in sarcoidosis: a systematic review. Curr Opin Pulm Med 2009; 15:499-506
12. Wirnsberger RM, de Vries J, Wouters EF, et al. Clinical presentation of sarcoidosis in The Netherlands an epidemiological study. Neth J Med 1998; 53:53-60
13. de Kleijn WP, Drent M, De Vries J. Nature of fatigue moderates depressive symptoms and anxiety in sarcoidosis. Br J Health Psychol 2013; 18:439-452
14. Chang B, Steimel J, Moller DR, et al. Depression in sarcoidosis. Am J Respir Crit Care Med 2001; 163:329-334
15. Elfferich MD, De Vries J, Drent M. Type D or 'distressed' personality in sarcoidosis and idiopathic pulmonary fibrosis. Sarcoidosis Vasc Diffuse Lung Dis 2011; 28:65-71
16. Demirkok SS, Basaranoglu M, Akinci ED, et al. Analysis of 275 patients with sarcoidosis over a 38 year period; a single-institution experience. Respir Med 2007; 101:1147-1154

Erratum corrige

In the issue 4-2013 of Sarcoidosis Vasculitis and Diffuse lung Diseases in the article “Role of Propionibacterium acnes in sarcoisosis: a meta-analysis “by Y. Zhou, Y. Hu, H. Li, The correct Corresponding Autor is: Huiping Li, MD E.mail: liw2013@126.com