

VCRC MANUAL OF OPERATIONS: THE DISEASE EXTENT INDEX (DEI)

I. For which entities can the DEI be applied?

The DEI was designed and has been validated for use in patients with *Wegener's granulomatosis*. The DEI can be applied to patients with active disease as well as patients with partial or complete remission. The DEI has also been partially validated in patients with *hepatitis C virus-associated cryoglobulinemic vasculitis*. Validation of the DEI for MPA is in progress.

II. The structure of the DEI Score sheet

The first column of the score sheet displays an abbreviated symbol which has been allocated to each organ system (e.g. "E" for ENT tract or "GI" for GI tract).

The second column lists all organ systems typically affected by small-vessel vasculitides.

The third column displays definitions of signs and symptoms typically attributable to active vasculitis. Manifestations not listed can also be scored if they are clearly attributable to *active* vasculitis.

The fourth column lists a set of diagnostic procedures which should be performed in order to diagnose, or alternatively rule out *active* vasculitis in the respective organ system. Diagnostic tests listed in brackets are not mandatory, but may be indicated to link certain clinical signs to active vasculitis and to rule out other causes like damage from previous vasculitis or infection.

The fifth column shows the score for each body system (score = 1 for constitutional symptoms, score = 2 for all other organ systems). Each body system is scored only once, irrespective of the number of affected organ within the organ system or the severity of the respective manifestation. For example, in a patient with bloody nasal discharge the score for "E" is 2. In a patient with tracheal stenosis, bloody nasal discharge and inner ear deafness, the score for "E" is also 2.

The sixth column contains boxes which should be ticked if active vasculitis is present in the respective body systems at the time of assessment. There is no distinction between new, worse or persistent disease. However, previous activity or damage is not counted. The scores for each of the ticked boxes (= affected organ systems) are added and give the total DEI score. For example, in a WG patient with bloody nasal discharge, tracheal stenosis, pulmonary nodules, arthritis and a weight loss > 10 % of body weight, the DEI score is 7 with the organ systems E, L, A and B being affected. Complete remission is defined by a DEI score of zero.

III. How to score an individual patient?

1. Check each organ system for disease attributable to *active* vasculitis
2. As a minimum requirement, use the diagnostic tests listed in the fourth column.
3. Use these tests also to rule out other causes of disease like damage or infection.
4. Tick the box in the last column, if any disease attributable to *active* vasculitis is present, irrespective if it is new, worse or persistent, mild or severe.
5. Do not tick the box if only disease resulting from damage, infection or other causes unrelated to *active* vasculitis are present.
6. Count the total score by adding the scores (listed in the fifth column) of all body systems with active vasculitis at the time of presentation.
7. In addition, abbreviations of the affected organ systems can be added in brackets in order to display which organ systems are affected (e.g. DEI 7 [E,L,A,B]).



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Subject ID: Clinic ID: Date form completed: / /

Day Mon Year

Investigator:

DEI Score Sheet

Sym bol	Organ Manifestation	Notes	Standard diagnostic procedure	Score	Present
E	ENT and upper airway	nasal obstruction, crusty, bloody discharge, non-infectious sinusitis, middle or inner ear deafness, laryngitis, tracheal stenosis	ENT exam (cranial MRI scan, biopsy, sinuscopy)	2	<input type="checkbox"/>
EY	inflammatory eye lesions	scleritis, episcleritis, retinitis, retroorbital mass, if not grown contiguously from the ENT tract	Ophthalmologic exam (MRI scan)	2	<input type="checkbox"/>
H	heart	pericardial effusion, new arrhythmias, myocardial infarction, myocarditis	ECG, chest x-ray, echocardiogram, myocardial scan, (cardiac catheter for potential proof of coronaritis and possible myocardial biopsy)	2	<input type="checkbox"/>
L	lung and lower airway	pulmonary hemorrhage, infiltrates, nodules; bronchial stenosis	chest x-ray, (high resolution CT bronchoscopy, biopsy)	2	<input type="checkbox"/>
K	kidney	nephritic sediment with or without deterioration of renal function	urinary sediment, serum creatinine, abdominal ultrasound (kidney biopsy)	2	<input type="checkbox"/>
GI	gastrointestinal tract	new bloody diarrhoea, histological proof of vasculitis	ultrasound (endoscopy w./wo. biopsy, angiography, surgery)	2	<input type="checkbox"/>
P	peripheral nervous system	motory and/or sensory distal symmetrical polyneuropathy or mononeuritis multiplex	neurological exam, ENG, EMG (MRI scan of muscles, sural nerve biopsy)	2	<input type="checkbox"/>
C	central nervous system	primary nodular CNS lesions, small vessel Vasculitis, white or gray matter	neurological exam, (EEG, cranial MRI scan, analysis of cerebrospinal fluid)	2	<input type="checkbox"/>
S	skin	palpable Purpura, digital gangrene, non-healing ulcerations	dermatologist's opinion, (skin biopsy)	2	<input type="checkbox"/>
A	arthralgias / arthritides	rheumatic complaints, including arthralgias, myalgias, non-destructive arthritides, myositis	x-ray, ultrasound, (bone scan, diagnostic joint puncture, muscle biopsy)	2	<input type="checkbox"/>
B	constitutional symptoms	fever > 38° C., weight loss > 10 % of body weight, fatigue, night sweats		1	<input type="checkbox"/>

Total Score

21

*MRI = magnetic resonance imaging, ECG = electrocardiography, EEG = electroencephalography, EMG = electromyography, ENG = electroneurography. Note: all symptoms in any organ system must be attributable to active vasculitis, other possible causes (especially damage, infection and/or malignoma) must be excluded. The column "notes" gives an idea about the most common organ specific signs and symptoms, but does not mean to be all-inclusive in any case. () in the last column = procedures that are optional, if standard procedures yield findings that need further specification.