Dear Colleagues,

Welcome to the first round of the DELPHI Survey of the Large-Vessel Vasculitis Sub Task Force.

This DELPHI Survey is a first step for developing preliminary consensual definitions of the characteristic elementary lesions observed by US in patients suffering from large-vessel vasculitis (LVV), which will subsequently be tested in future US exercises. Consensus on each recommendation will be achieved if the agreement exceeds 75%.

This DELPHI survey incorporates some of the already published statements and recommendations.

Section A: Definition of normal and arteriosclerotic US vascular appearance
Section B: Definition of pathology
Section C: Consensus on how to perform ultrasound in suspected LVV

If you have decided on your degree of agreement, please underline the respective number, (e.g. 5, if you strongly agree).

Best regards,
Wolfgang Schmidt, Christina Duftner and Christian Dejaco
Section A: Consensus on definitions of normal and arteriosclerotic ultrasound appearance of arteries

Please indicate your level of agreement with each statement.

1. Definition of US appearance of normal temporal arteries:

Pulsating, compressible artery with anechoic lumen surrounded by mid- to hyperechoic tissue. Using US equipment with high resolution, the intima-media complex presenting as a homogenous, hypo- or anechoic echostructure delineated by two parallel hyperechoic margins (“double line pattern”) may be visible.

How do you agree with this sentence? (please underline)

(strongly disagree) 1 2 3 4 5 (strongly agree)

Additional comments or suggestions (optional):

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2. Definition of US appearance of normal extra-cranial large arteries (e.g. carotid, vertebral, subclavian, axillary, femoral and popliteal arteries):

Pulsating, hardly compressible artery with anechoic lumen; the intima-media complex presents as a homogenous, hypo- or anechoic echostructure delineated by two parallel hyperechoic margins (“double line pattern”), which is surrounded by mid- to hyperechoic tissue.

How do you agree with this sentence? (please underline)

(strongly disagree) 1 2 3 4 5 (strongly agree)

Additional comments or suggestions (optional):

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3. Definition of US appearance of arteriosclerotic arteries:

Non-homogenous and in part hyperechoic, irregularly delineated, eccentric vessel wall alteration.

How do you agree with this sentence? (please underline)

(strongly disagree)  1  2  3  4  5  (strongly agree)

Additional comments or suggestions (optional):

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Section B: Consensus on definitions of pathologic ultrasound findings in arteries

This section will ask the group’s opinion on the candidate elementary lesions (previously identified by the literature review).

Definition of US elementary lesions of vasculitis

Vasculitis can be characterized by the following elementary lesions on US.

Please indicate your level of agreement with each statement.

1. Definition of US appearance of vasculitis – “HALO SIGN”:

   a) Circumferential, homogenous, hypoechoic wall thickening, well delineated towards the luminal side, visible both in longitudinal and transverse planes.

   How do you agree with this sentence? (please underline)

   (strongly disagree)  1  2  3  4  5  (strongly agree)

   Additional comments or suggestions (optional):

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   b) Circumferential, homogenous, hypoechoic wall thickening, well delineated towards the luminal side, visible both in longitudinal and transverse planes, which needs to exceed a defined cut-off value as defined below for the involved artery/arteries.

   How do you agree with this sentence? (please underline)

   (strongly disagree)  1  2  3  4  5  (strongly agree)

   Additional comments or suggestions (optional):

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1.1. Inclusion of a cut-off value for the definition of the “halo sign” in US?

In the current literature, cut-off values for the definition of the “halo sign” were mostly proposed for temporal arteries, whereas for other (particularly extra-cranial large) arteries, there is little evidence for a valuable cut-off.

For which arteries is the inclusion of a cut-off value for the definition of the “halo sign” useful? Please indicate your response in Table 1 and propose a cut-off providing (in your view) optimal sensitivity and specificity for diagnosis of large vessel vasculitis.

Table 1. Usefulness of cut-off values for definition of the “halo sign” for temporal and other extra-cranial large arteries.

<table>
<thead>
<tr>
<th>VESSEL</th>
<th>Inclusion of cut-off value (please indicate your suggestion)</th>
<th>Inclusion of cut-off value is NOT useful</th>
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<tbody>
<tr>
<td>temporal arteries</td>
<td>≥0.3mm</td>
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<tr>
<td></td>
<td>≥0.4mm</td>
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<tr>
<td></td>
<td>≥0.5mm</td>
<td></td>
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<tr>
<td></td>
<td>≥0.7mm</td>
<td></td>
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<td></td>
<td>≥1.0mm</td>
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<tr>
<td></td>
<td>other value:</td>
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<tr>
<td>axillary arteries</td>
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<td>common carotid arteries</td>
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<td>vertebral arteries</td>
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<tr>
<td>subclavian arteries</td>
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<tr>
<td>facial arteries</td>
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<tr>
<td>occipital arteries</td>
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<td>abdominal aorta</td>
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<tr>
<td>superficial femoral arteries</td>
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<td>popliteal arteries</td>
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</table>

Additional comments or suggestions (optional):

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2.1. Definition of US appearance of vasculitis – STENOSIS of temporal arteries:

a) A stenosis is characterized by aliasing and persistent diastolic flow by colour Doppler US. The maximum systolic flow velocity determined within the stenosis by pulsed wave (pw)-Doppler ultrasound is >2 times higher than the flow velocity before or behind the stenosis.

How do you agree with this sentence? (please underline)
(strongly disagree) 1 2 3 4 5 (strongly agree)

Additional comments or suggestions (optional):
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b) A segmental, ≥2-fold increase of the peak systolic velocity of blood flow accompanied by poststenotic turbulence not attributable to other reasons.

How do you agree with this sentence? (please underline)
(strongly disagree) 1 2 3 4 5 (strongly agree)

Additional comments or suggestions (optional):
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c) Increase of blood flow velocity being more than twice the rate recorded in the area before the stenosis, perhaps with wave forms demonstrating turbulence and reduced velocity behind the area of stenosis.

How do you agree with this sentence? (please underline)
(strongly disagree) 1 2 3 4 5 (strongly agree)

Additional comments or suggestions (optional):
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2.2. Definition of US appearance of vasculitis – STENOSIS of other extra-cranial large arteries:

a) Typical vasculitic vessel wall thickening together with characteristic Doppler curves showing turbulences and increased systolic and diastolic blood flow velocities.

How do you agree with this sentence? (please underline)

(strongly disagree) 1 2 3 4 5 (strongly agree)

Additional comments or suggestions (optional):

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b) A reduction of the artery lumen of more than 50% of the original lumen because of typical vasculitic vessel wall thickening together with characteristic Doppler curves showing turbulences and increased systolic and diastolic blood flow velocities.

How do you agree with this sentence? (please underline)

(strongly disagree) 1 2 3 4 5 (strongly agree)

Additional comments or suggestions (optional):

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Cc) Typical vasculitic wall swelling together with pansystolic spectral broadening with a peak systolic velocity of >125 cm/s.

How do you agree with this sentence? (please underline)

(strongly disagree) 1 2 3 4 5 (strongly agree)

Additional comments or suggestions (optional):

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3. Definition of US appearance of vasculitis – OCCLUSION:

a) Absent colour flow in a clearly visible vessel on colour Doppler US.

How do you agree with this sentence? (please underline)

(strongly disagree) 1 2 3 4 5 (strongly agree)

Additional comments or suggestions (optional):

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b) Unability of US to delineate colour in the former arterial lumen which shows a hypoechoic or mid-echoic appearance.

How do you agree with this sentence? (please underline)

(strongly disagree) 1 2 3 4 5 (strongly agree)

Additional comments or suggestions (optional):

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c) Absence of colour Doppler signals in a visible artery filled with hypoechoic material, even with low pulse repetition frequency (PRF) and high colour gain.

How do you agree with this sentence? (please underline)

(strongly disagree) 1 2 3 4 5 (strongly agree)

Additional comments or suggestions (optional):

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4. Definition of US appearance of vasculitis – COMPRESSION SIGN of temporal arteries:

a) The thickened arterial wall remains visible upon compression, i.e. the echogenicity contrasts differently than the surrounding tissue due to vasculitic vessel wall thickening.

How do you agree with this sentence? (please underline)

(strongly disagree) 1 2 3 4 5 (strongly agree)

Additional comments or suggestions (optional):

b) The thickened arterial wall remains visible upon compression as hypoechoic material compared to the surrounding tissue.

How do you agree with this sentence? (please underline)

(strongly disagree) 1 2 3 4 5 (strongly agree)

Additional comments or suggestions (optional):

c) The thickened arterial wall remains visible upon compression, i.e. the echogenicity contrasts hypoechogenic due to vasculitic vessel wall thickening in comparison to the mid- to hyperechogenic surrounding tissue.

How do you agree with this sentence? (please underline)

(strongly disagree) 1 2 3 4 5 (strongly agree)

Additional comments or suggestions (optional):
5. Definition of US appearance of vasculitis – VESSEL WALL PULSATION of temporal arteries:

a) Absent vessel wall pulsation.

How do you agree with this sentence? (please underline)

( strongly disagree) 1 2 3 4 5 ( strongly agree)

Additional comments or suggestions (optional):

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6. Which US elementary lesions are important for the ultrasound definition of vasculitis

6.1. Vasculitis can be defined by the presence of the following pathological lesions

Please mark your response in the table below.

<table>
<thead>
<tr>
<th>ELEMENT</th>
<th>YES</th>
<th>If YES – must it be absolutely present for defining VASCULITIS</th>
<th>NO</th>
</tr>
</thead>
<tbody>
<tr>
<td>Halo sign</td>
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<td></td>
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<tr>
<td>Stenosis</td>
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<td></td>
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<tr>
<td>Occlusion</td>
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<tr>
<td>Compression sign (temporal arteries)</td>
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<tr>
<td>Lack of vessel wall pulsation (temporal arteries)</td>
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</table>

6.2. Which elements above need to be present to meet the MINIMUM requirement for vasculitis in your opinion? Please underline!

Elements: 1 (Halo) 2 (Stenosis) 3 (Occlusion) 4 (Compression sign) 5 (Lack of vessel wall pulsation)

Additional comments or suggestions (optional):

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General considerations for the standardization of US in diagnosis of LVV.

Please indicate your level of agreement with each statement.

**Definitions on the performance of measurements in US for the assessment of vasculitis**

1. **Assessment of the “halo sign” by ultrasound:**

   a) The typical vessel wall thickening of the “halo sign” should be assessed by the “interface to interface” method (inner layer of the luminal-intimal margin to the inner layer of the medial-adventitial margin perpendicularly) at the site with the greatest thickness of the intima-media complex.

   How do you agree with this sentence? (please underline)

   (strongly disagree) 1 2 3 4 5 (strongly agree)

   Additional comments or suggestions (optional):
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   b) The typical vessel wall thickening of the “halo sign” should be assessed by the “interface to interface” method (inner layer of the luminal-intimal margin to the inner layer of the medial-adventitial margin perpendicularly) at the site with the greatest thickness of the intima-media complex, given as average of least 3 measurements.

   How do you agree with this sentence? (please underline)

   (strongly disagree) 1 2 3 4 5 (strongly agree)

   Additional comments or suggestions (optional):
   ..................................................................................................................................................................................
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c) The typical vessel wall thickening of the “halo sign” should be assessed by the “interface to interface” method (inner layer of the luminal-intimal margin to the inner layer of the medial-adventitial margin perpendicularly) at the site with the greatest thickness of the intima-media complex in a longitudinal plane.

How do you agree with this sentence? (please underline)

(strongly disagree) 1 2 3 4 5 (strongly agree)

Additional comments or suggestions (optional):
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d) The typical vessel wall thickening of the “halo sign” should be assessed by the “interface to interface” method (inner layer of the luminal-intimal margin to the inner layer of the medial-adventitial margin perpendicularly) at the site with the greatest thickness of the intima-media complex in a longitudinal plane at the distal vessel wall.

How do you agree with this sentence? (please underline)

(strongly disagree) 1 2 3 4 5 (strongly agree)

Additional comments or suggestions (optional):
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2. **Assessment of the “compression sign” by ultrasound:**

a) The compression sign should be assessed by applying pressure via the transducer until the lumen of the temporal artery occludes and no arterial pulsation remains visible.

**How do you agree with this sentence? (please underline)**

(strongly disagree) 1 2 3 4 5 (strongly agree)

Additional comments or suggestions (optional):

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3. **Assessment of the “vessel wall pulsation” by ultrasound:**

a) The vessel wall pulsation is assessed by the M-mode of the far wall of the temporal artery at non-affected sites and sites with suspicion of vasculitis vessel wall thickening.

**How do you agree with this sentence? (please underline)**

(strongly disagree) 1 2 3 4 5 (strongly agree)

Additional comments or suggestions (optional):

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b) The vessel wall pulsation is assessed subjectively by B-mode.

**How do you agree with this sentence? (please underline)**

(strongly disagree) 1 2 3 4 5 (strongly agree)

Additional comments or suggestions (optional):

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Thank you very much!