Ultrasound in Large Vessel Vasculitis

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Basics of Vascular Ultrasound

The Outcome Measures in Rheumatology (OMERACT) group on ultrasound in large vessel vasculitides (LVV) has developed definitions and standardized the ultrasonographic findings of inflammation in temporal and axillary arteritis [3]. Cut-offs for the normal thickness in cranial and axillary arteries have been developed, mainly to distinguish vasculitis from atherosclerosis (Table 1) [4].

We suggest the examination of the temporal artery in two planes when the halo sign appears to be present. Compression sign should always apply in short axis to avoid false negatives (the inflamed artery can slide to the side if compressed in the long axis). For the large vessels, examination in long axis is sufficient.

The Fast Track ultrasound GCA clinics reduce significantly the number of patients suffering from irreversible visual loss [5, 6] and have been introduced in many countries thus improving outcomes for GCA patients.

The anteromedial ultrasound examination technique which is demonstrated in this chapter includes a complete and systematic examination of the supra-aortic tree, the aortic arch, the ascending and the abdominal aorta in all LVV patients (Table 2). Preliminary results have demonstrated the superiority of the anteromedial ultrasound examination in the identification of large-vessel involvement in GCA patients (up to 79% of all GCA patients) [7]. Additionally, we use the anteromedial ultrasound approach as a diagnostic and follow-up tool for all GCA and TAK patients. Furthermore, ultrasound is used to confirm a flare or to evaluate response to treatment (Table 2). The anteromedial approach requires a recording

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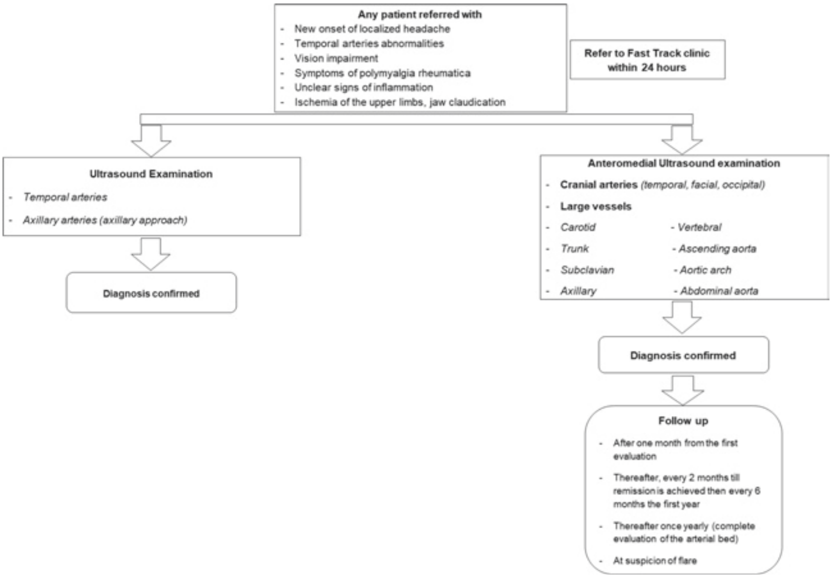
Table 1 Overview of the cut-off values for the cranial and axillary arteries [4, 19]

Artery	Cut-off values (mm)
Temporal artery common superficial	0.42
Temporal artery frontal branch	0.34
Temporal artery parietal branch	0.29
Facial artery	0.37
Axillary artery	1.00

Table 2 Flowchart of the suggested ultrasound evaluation of LVV patients



of the vasculitic changes and a comparison between consequent examinations to identify differences in IMT (for the response to treatment, damage or disease flare) [8].

Definitions

Intima media thickness (IMT) is the "area of tissue starting at the luminal edge of the artery and ending at the boundary between the media and the adventitia" [9]. Any transducer > 15 MHz for the cranial arteries and > 10 MHz for the supra-aortic arteries should be sufficient for the proper visualization of the arteries [1, 10]. To examine the aorta, a phased array transducer for the ascending aorta and the aortic arch and a curvilinear transducer for the abdominal aorta and the deep pelvic vessels are considered sufficient [1, 10].

OMERACT Ultrasound Working Group in large vessel vasculitis standardized the definitions of vasculitic vessel wall changes (halo sign) and compression sign and tested the reliability of the developed definitions [3, 11]. OMERACT has defined the ‘halo sign’ as ‘’a homogeneous, hypoechoic wall thickening well-delineated towards the luminal side visible both in longitudinal and transverse planes, most commonly concentric in transverse planes” [3]. The compression sign helps to distinguish inflammation of the vessel wall in cranial arteries from pseudo halo due to incorrect ultrasound equipment adjustments or poor technique [12]. According to the OMERACT definition, ‘’The thickened arterial wall remains visible upon compression, i.e. the echogenicity contrasts hypoechoic due to vasculitis vessel wall thickening in comparison to the mid-to hyperechoic surrounding tissue’’ [3] (Fig. 21c).

The use of cut-off values for IMT of the cranial and axillary arteries may be useful to distinguish vasculitis from normal arteries in patients suspected to have GCA [4]. The cut-off values are presented in Table 1.

Atherosclerosis which is one of the main challenges in ultrasound in vasculitis is defined as hypo-, iso- or hyperechoic, non-homogeneous and localized plaques seen mainly in the large vessels at bifurcations. In some patients, atherosclerosis may present as an iso- or hyperechoic, homogeneous wall thickening thus making difficult the distinction from chronic vasculitic changes (Fig. 23). In rare cases in the cranial vessels, a biopsy may be necessary to distinguish vasculitis from atherosclerosis.

It is important to note that halo sign is not a distinct feature of GCA, but can appear in other vasculitides (ANCA associated vasculitides, polyarteritis nodosa), infections, or other rare conditions [13–15].

Ultrasonographically, there are no differences in echogenicity in TAK patients compared to GCA patients. The slope sign is a pathologically increased IMT that spreads over a long arterial segment and slides down to a normal brachial artery and is mainly observed in GCA patients [16, 17] (Fig. 29b).

Observational cohort studies demonstrated the distinct differences regarding the distribution of vasculitic involvement among GCA and TAK. GCA involves predominantly the cranial, subclavian and axillary arteries, while TAK involves the left carotid, the left subclavian and the abdominal arteries [18].

Settings and Pitfalls

Gray Scale

Now, moving to the ultrasound examination of the arterial system, the depth of the image should be kept at 1–1.5 cm for the temporal artery and at 2–2.5 for the facial and occipital arteries. For the supraaortic arteries, the depth should be sufficient to visualize the artery in the middle of the screen. Brightness should be enough to visualize the vessel wall and can be adjusted by the B mode gain. An image that is too bright, may lead to the incorrect conclusion that the increased IMT is atherosclerotic while too dark, images may miss the inflammatory changes of the vessel wall. Frequency should be adjusted according to the depth in which the vessel is localized. Superficial vessels require higher frequencies, while deeper vessels lower frequencies.

Color Doppler

Color Doppler has the advantage of visualizing the blood flow direction thus improving the diagnostic capability of ultrasound especially if stenosis and occlusion are present. Color Doppler is preferred for the visualization of the cranial arteries and if stenosis or occlusion are suspected in the large vessels. In Color Doppler the angle of insonation of the Doppler window should be preferably kept between 30 and 60 degrees and in any case < 90 degrees to avoid inadequate filling of the arterial lumen. In addition, the color Doppler gain should be adjusted so the color fills the lumen completely (Fig. 1).

Pulse Repetition Frequency (PRF) is one of the important adjustments in vascular ultrasound. PRF is the number of ultrasound pulses emitted by the transducer over a designated period of time. For the examination of the cranial arteries, a PRF of 2–3 MHz is sufficient while for the large vessels a higher value of 4 MHz is adequate.

Pitfalls

Blooming Appears when the color exceeds beyond the artery walls. The reason for blooming is that the color gain is too high or PRF is too low (Fig. 21).
Solution: Adjust the color gain and PRF accordingly.

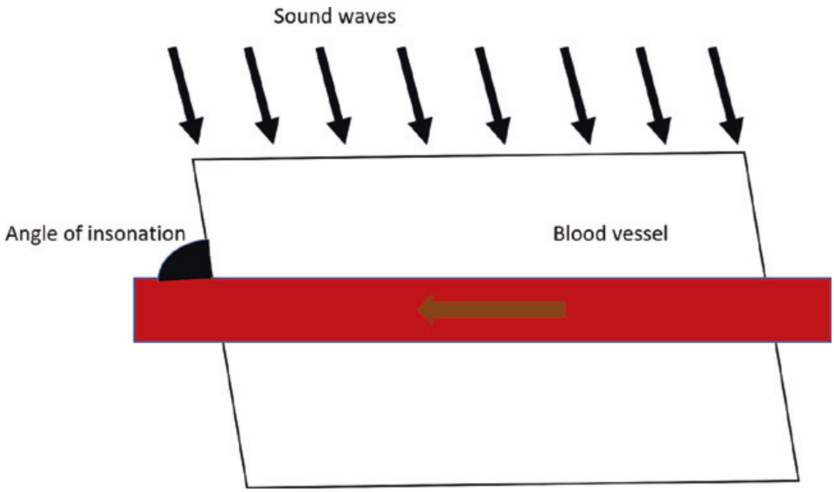


Fig. 1 Angle of insonation

Pseudo halo. In pseudo halo, the thickened area appears anechoic and this should alert the ultrasonographer of the possibility of wrong adjustments as vasculitis appears hypoechoic and fluid (in this case the blood flow) anechoic (Fig. 22). Pseudo halo appears if the color gain is low, the PRF is very high or if the Color Box is not steered properly. Pseudo halo is also observed in areas of hair such as the scalp especially in the parietal area.

Solution: Raise the gain, reduce the PRF to approximately 2. 5 kHz or steer the color box appropriately in order for the sound waves to meet the blood flow at an angle between 30 and 60 degrees (Fig. 1). In hairy areas use copious amounts of gel. However, the most practical solution to avoid pseudo halo is to compress the artery. In the case of a positive compression sign, the residual hypoechoic tissue after the compression is the thickened inflamed arterial wall (Fig. 24c).

Ultrasonographic Atlas of Examination of the Cranial Arteries, the Supraaortic Arteries and the Aorta

A. **Cranial Arteries**

See Fig. 2.

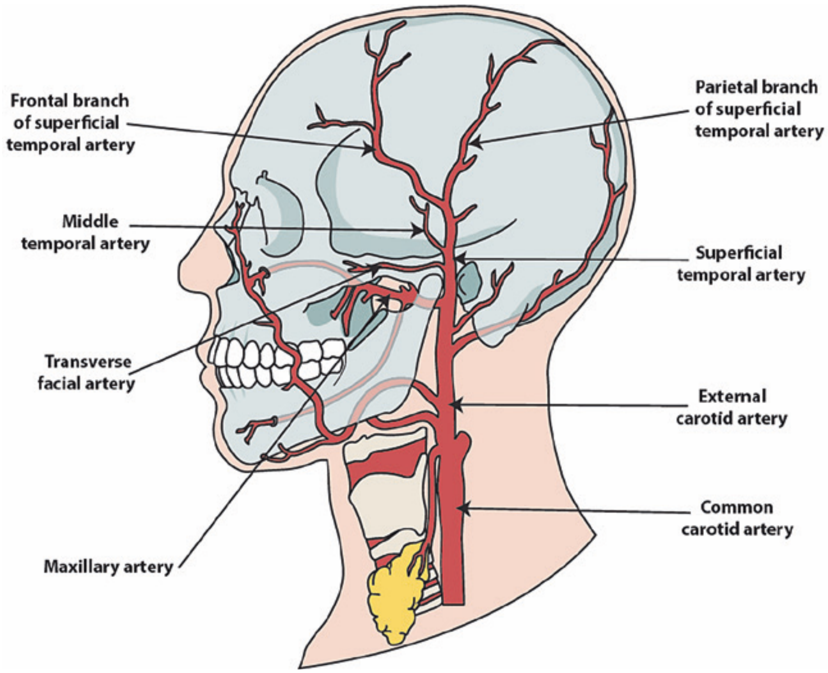


Fig. 2 Anatomical overview of the cranial vessels

Temporal Artery

The examination of the temporal arteries begins with the transducer longitudinally in front of the ear (Fig. 3a,b). It continues towards the parietal branch (Fig. 4a,b) until the top of the head and then turns transversely to the bifurcation. The frontal artery should be identified longitudinally and followed up to the frontal area (Fig. 5a,b) and then transversely backward to the bifurcation. The examination ends with a transverse view of the common branch.

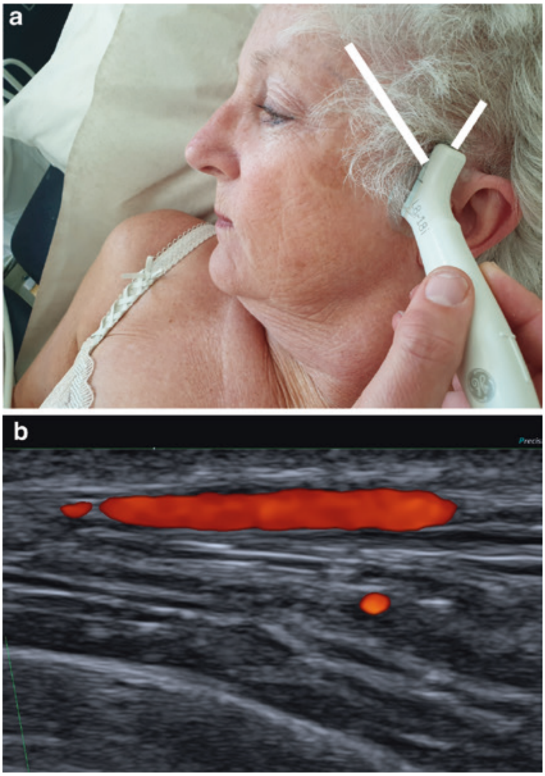


Fig. 3 Examination of the **common temporal artery**: **a** position of the transducer, **b** ultrasound image, longitudinal view

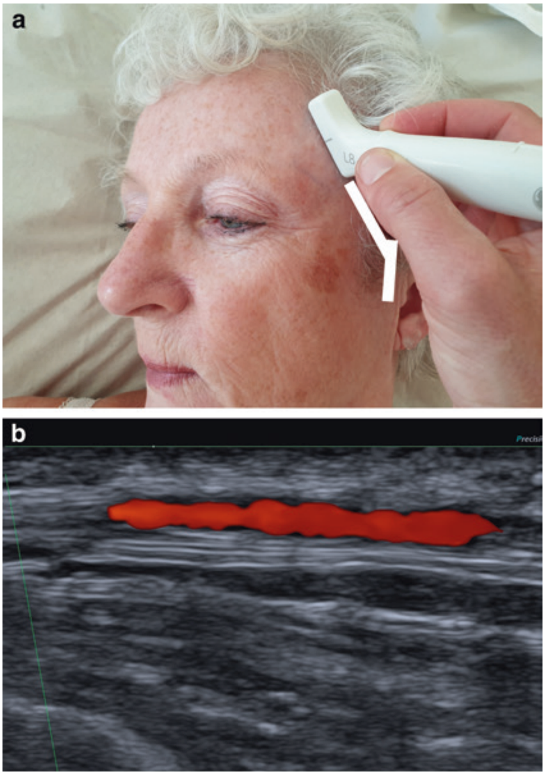


Fig. 4 Examination of the **frontal temporal artery**: **a** recommended position of the transducer **b** ultrasound image, longitudinal view

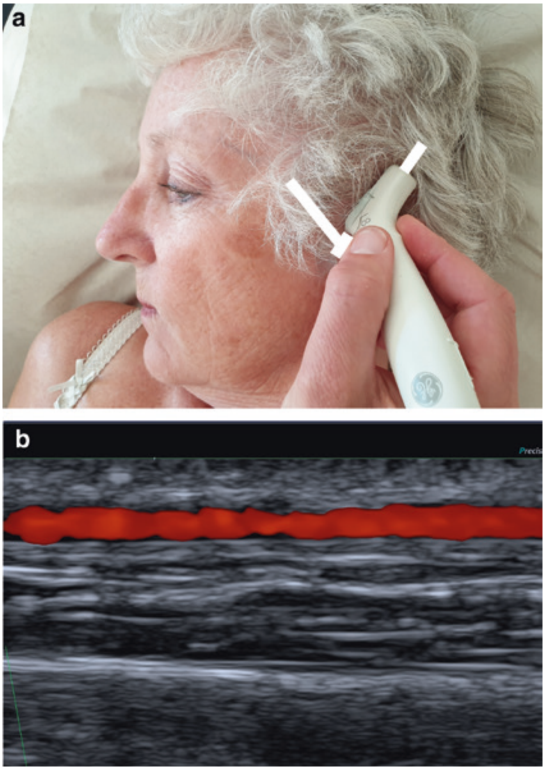


Fig. 5 Examination of the **parietal branch** of the temporal artery: **a** recommended position of the transducer, **b** ultrasound image, longitudinal view

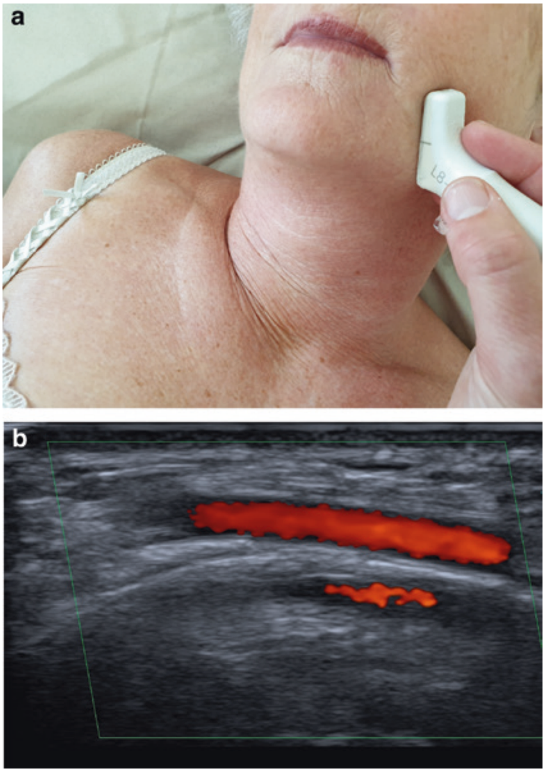


Fig. 6 Examination of the **facial artery**: **a** recommended position of the transducer **b** ultrasound image, longitudinal view

Distal Facial Artery

The examination starts with the identification of the facial artery in a transverse view in the middle of the jaw and continuing proximally towards the ear (Fig. 6).

Occipital Artery

The occipital artery is identified just below the mastoid process (Fig. 7) and should be followed longitudinally towards the back of the head.

B. **Ultrasonographic Examination of the Supraaortic Arteries**

The examination of the supraaortic arteries requires the use of medium frequency probes to visualize the deep vessels (e.g. subclavian, vertebral). The phased array probe should be used to examine the ascending aorta, the aortic arch, and the

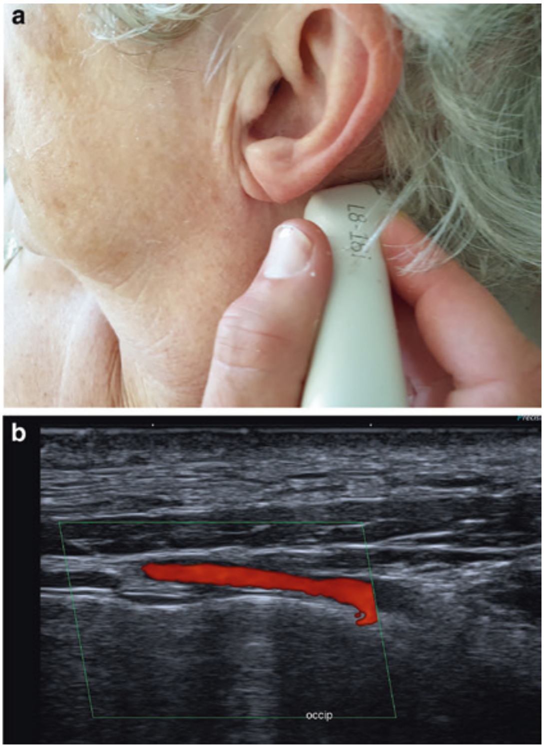


Fig. 7 Examination of the **occipital artery**: **a** recommended position of the transducer **b** ultrasound image, longitudinal view

aortic valve. The low-frequency curvilinear probe is an excellent choice for the visualization of the abdominal aorta and the deep pelvic arteries.

The examination starts by visualizing the carotid artery in the short axis in the middle of the neck. An important point here is the identification of the internal and external carotid artery and the use of Doppler to assure that blood flow is present.

Anatomically, the axillary artery is divided into 3 parts according to its relation to the pectoralis minor muscle (first part proximal or suprapectoral, second part behind or subscapular and the third part or infrapectoral distal to pectoralis minor).

However, for practical reasons, we divide ultrasonographically the axillary artery into two segments with the subscapular artery being the turning point: one proximal (from clavicle to the subscapular artery) and one distal (from subscapular to the deep brachial artery) (Figs. 8 and 9).

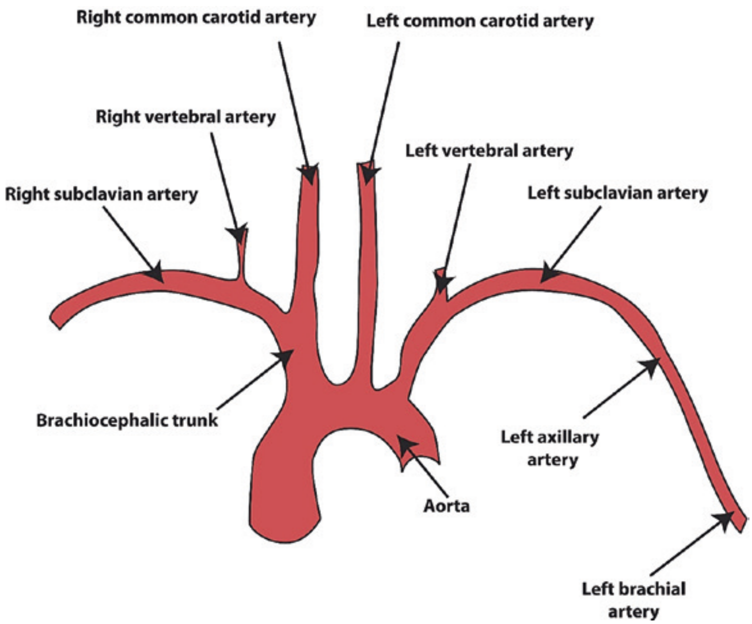


Fig. 8 Anatomical overview of the supraaortic arteries and aorta

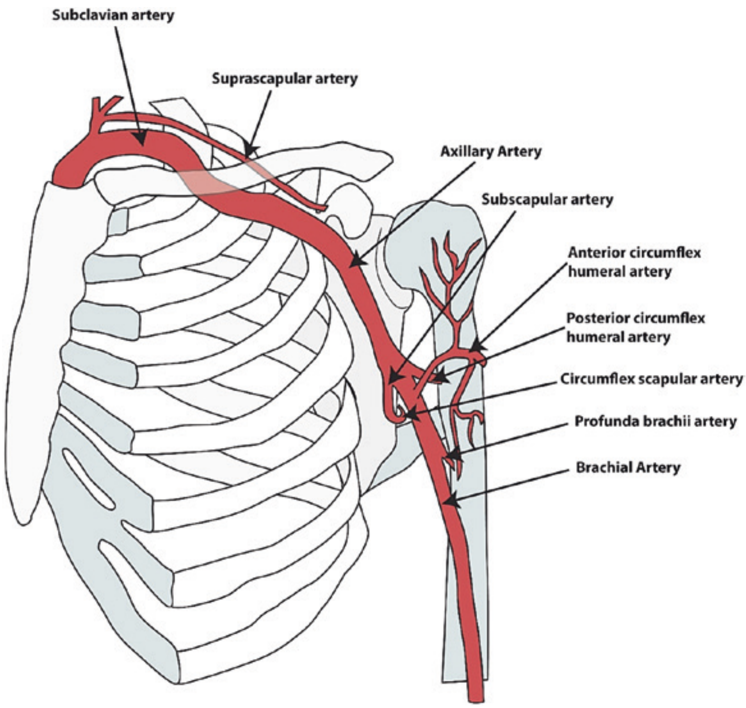


Fig. 9 Anatomical overview of supraaortic arteries, left side

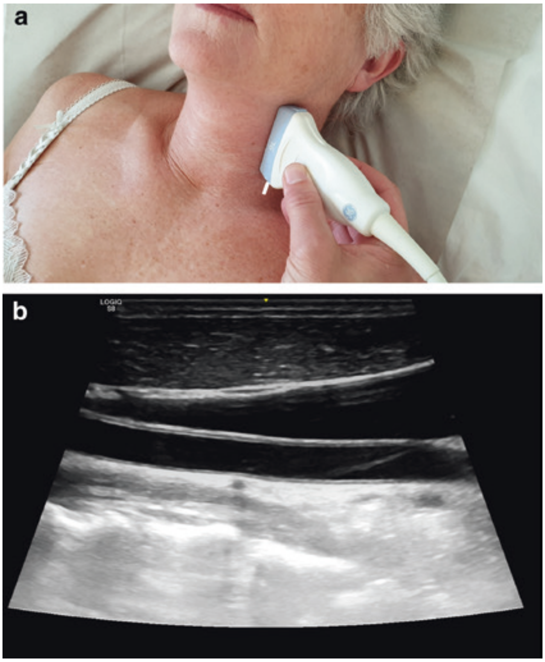


Fig. 10 Examination of the **carotid artery**: **a** recommended transducer position, **b** ultrasonographic image, longitudinal view

Carotid Artery

The examination in the middle of the neck just above the clavicle with the transducer placed transversely. The artery is followed in its whole length both proximally to the clavicle and distally to the skull base. Internal and external arteries are also identified (Fig. 10).

Vertebral Artery

The vertebral artery is localized posteriorly and lateral to the carotid artery. Visualization of the vertebral artery requires the identification of the carotid artery in a longitudinal view. Subsequently, the transducer should be slightly pointed laterally until the vertebral artery is identified (Fig. 11). Color Doppler should be used to identify the artery. The color in the vertebral artery should be compared to the color of the carotid artery to rule out subclavian steal syndrome.

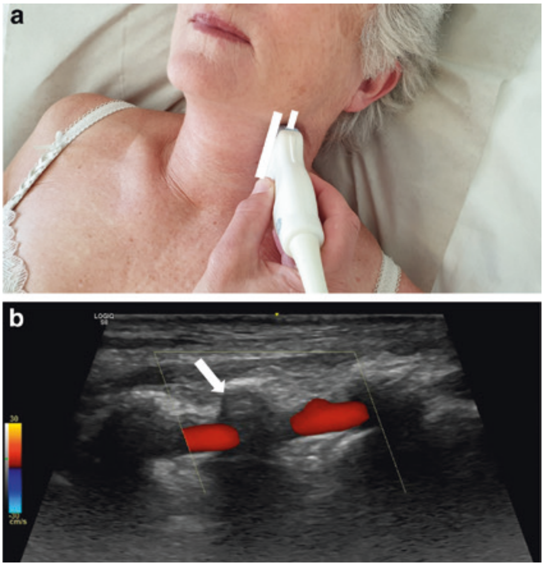


Fig. 11 Examination of the **vertebral artery**: **a** recommended transducer position, **b** ultrasonographic view of the vertebral artery, passing through the vertebra (white arrow)

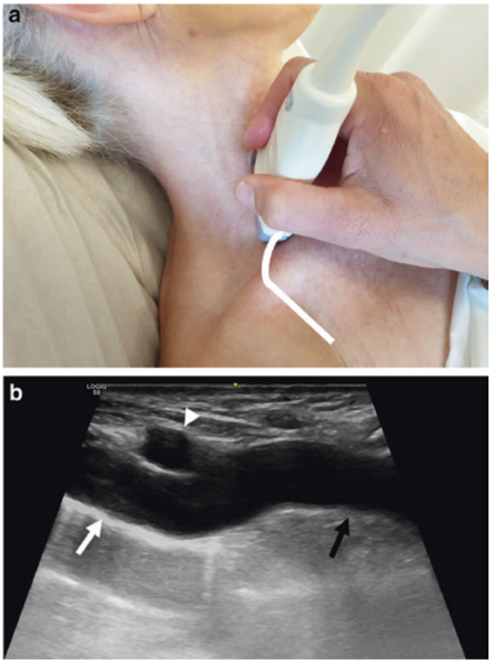


Fig. 12 Examination of the **right subclavian artery**: **a** recommended transducer position, **b** ultrasonographic view of the brachiocephalic trunk (black arrow), carotid artery (arrowhead), subclavian artery (white arrow)

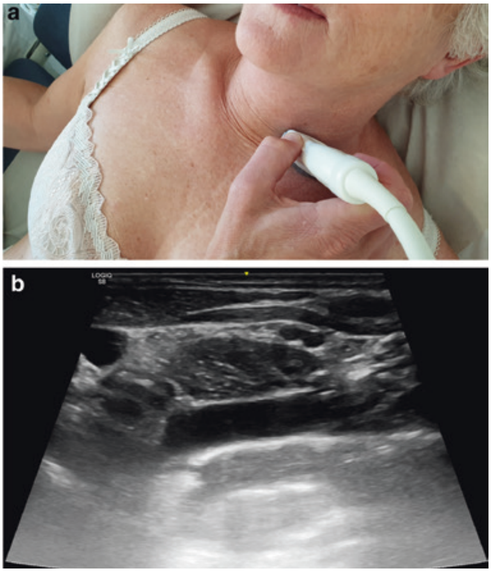


Fig. 13 Examination of the **left subclavian artery**: **a** recommended transducer position, **b** ultrasonographic view of the left subclavian artery

Brachiocephalic Trunk and Subclavian Artery

The carotid artery is followed with the transducer proximally in a transverse plane. When the transducer is in contact with the clavicle orientated slightly towards the lung the brachiocephalic trunk arises longitudinally at the right side (Fig. 12) and the subclavian artery at the left side (Fig. 13). The subclavian artery should be followed distally to the proximal edge of the clavicle.

Subclavian Artery

See Fig. 13.

Proximal Axillary Artery

The examination starts with the distal part of the subclavian artery visualized and the transducer in contact with the bony area of the clavicle, the transducer should be moved over the clavicle and placed in the sulcus between the deltoid and the pectoralis muscles (Fig. 14) until the subscapular artery which appears at the lower arterial wall (Fig. 14b).

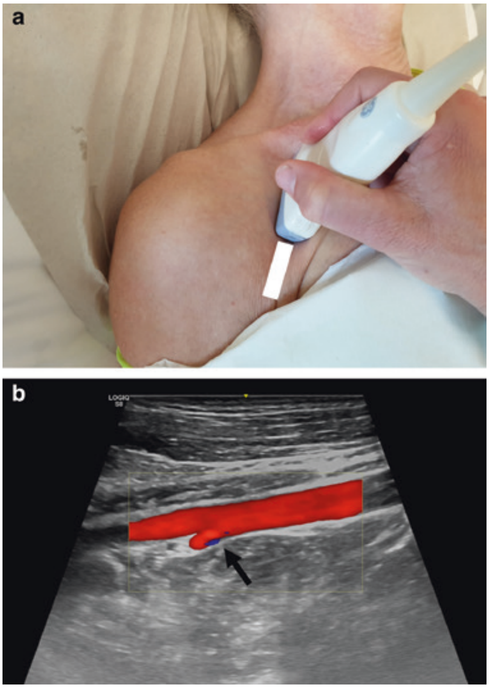


Fig. 14 Examination of the proximal axillary artery: **a** recommended transducer position, **b** ultrasonographic view (subscapular artery-black arrow)

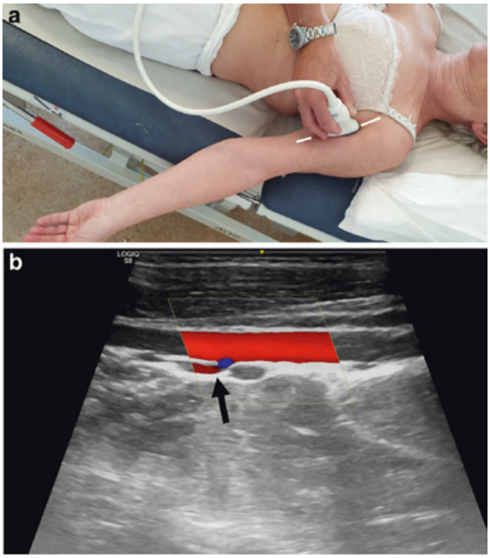


Fig. 15 Examination of the **distal axillary artery**: **a** recommended transducer position, anteromedial approach **b** ultrasonographic view of the distal axillary and brachial artery (including the deep brachial artery-black arrow)

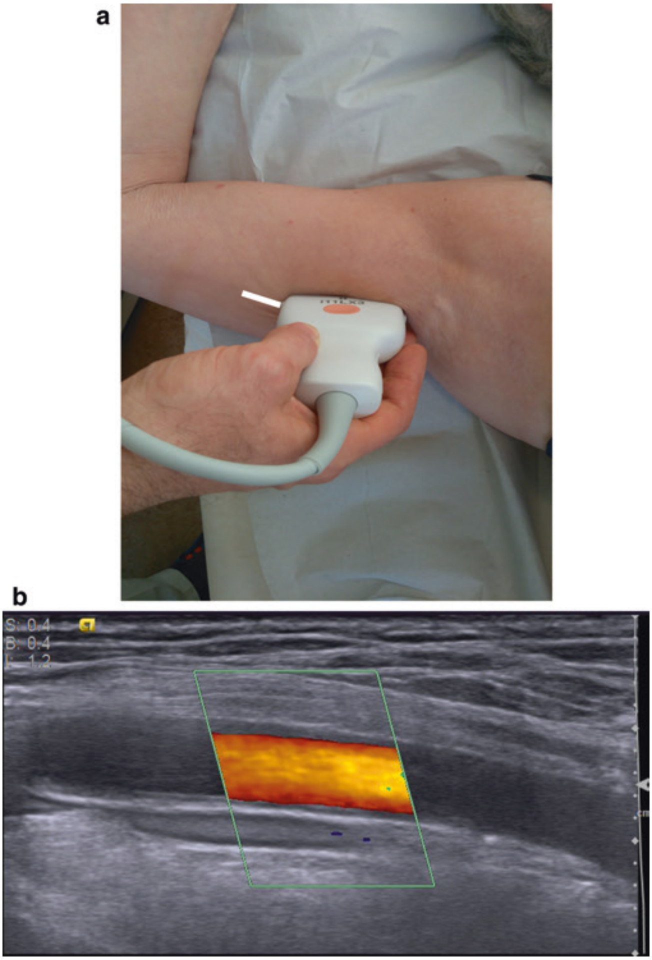


Fig. 16 Examination of the **distal axillary artery,** axillary approach: **a** recommended transducer position, **b** ultrasonographic view of the distal axillary and brachial artery (including the deep brachial artery-black arrow)

Distal Axillary Artery

The distal axillary artery appears after the subscapular artery, by moving the transducer anteromedially (Fig. 15). The artery is visualized through the biceps muscle and appears deeper compared to the axillary scanning. Both the upper and lower vessel wall are pictured. The axillary approach allows the demonstration of the axillary artery with great detail due to the superficial position of the axillary artery. The transducer is placed between the biceps and triceps muscles longitudinally (Fig. 16). However, because of the reverberation artifacts, the upper vessel wall is

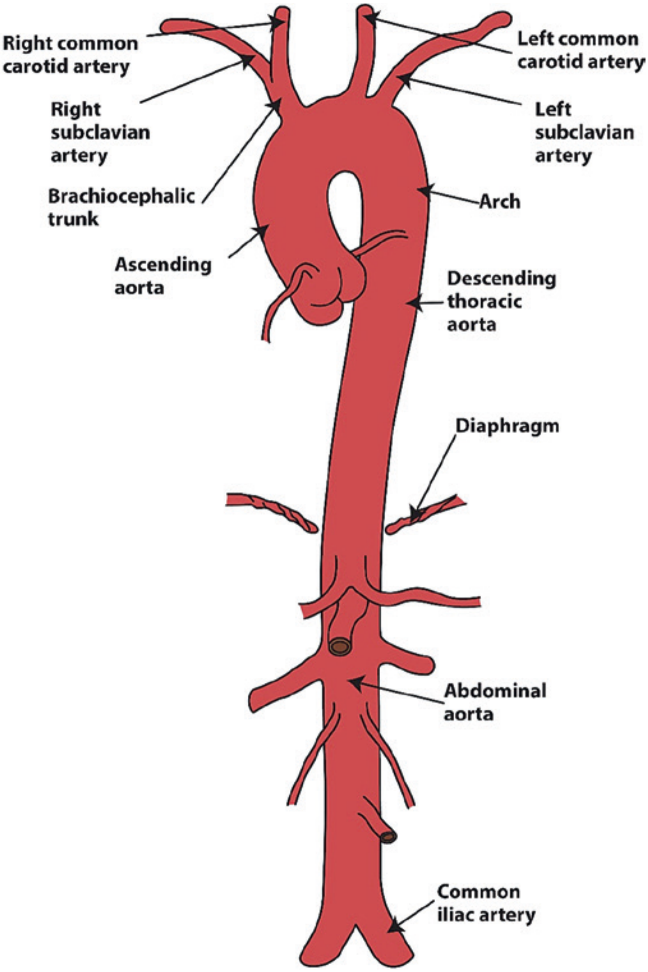


Fig. 17 Overview of the aortic tree

not visible, and the use of color Doppler is strongly advised to identify vasculitic involvement of the upper wall (Fig. 17).

C. **Aorta**

The visualization of the aorta requires the use of lower frequency transducers. The ascending aorta can be visualized by using a phased array transducer in the left parasternal longitudinal view (Fig. 18) and the aortic arch in the suprasternal notch view (Fig. 19). To examine the abdominal aorta the use of a curvilinear low-frequency transducer is advised. The abdominal aorta is identified by using the subcostal approach (Fig. 20) in a transverse view and then switching to a longitudinal view by moving the transducer towards the umbilicus. The bifurcation with both

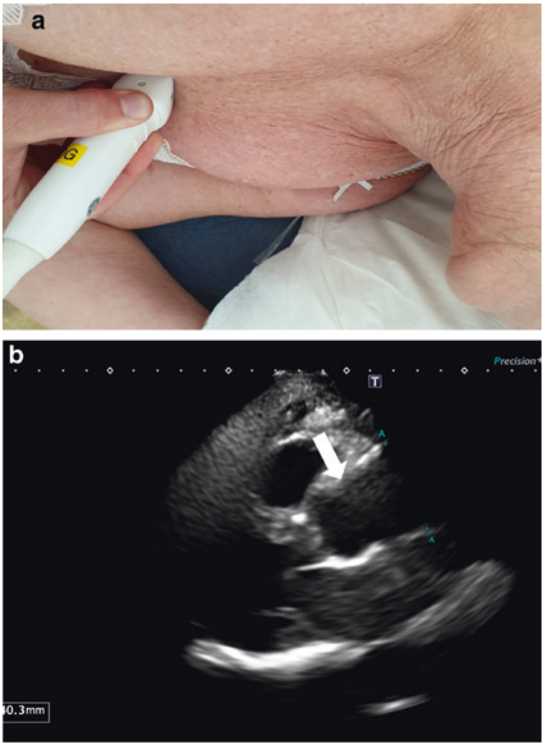


Fig. 18 Examination of the ascending aorta and the aortic valve: **a** recommended transducer position, **b** ultrasonographic view of the ascending aorta (white arrow)

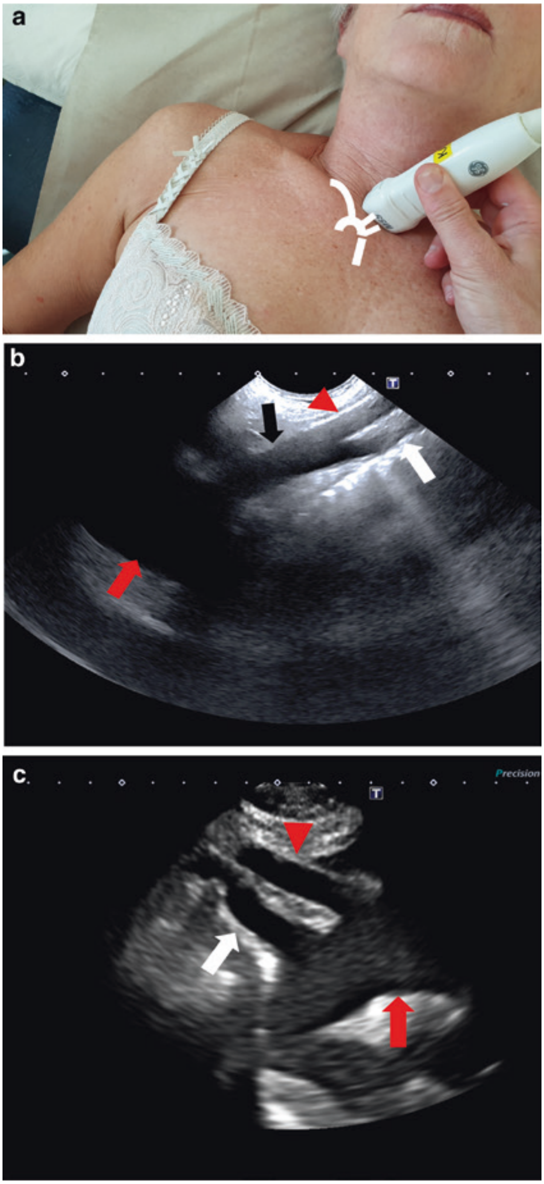


Fig. 19 Examination of the **aortic arch**: **a** recommended transducer position, **b** ultrasonographic view of the brachiocephalic trunk (black arrow), carotid artery (red arrowhead), subclavian artery (white arrow), aorta (red arrow), right side **c** carotid and subclavian artery, aorta, left side

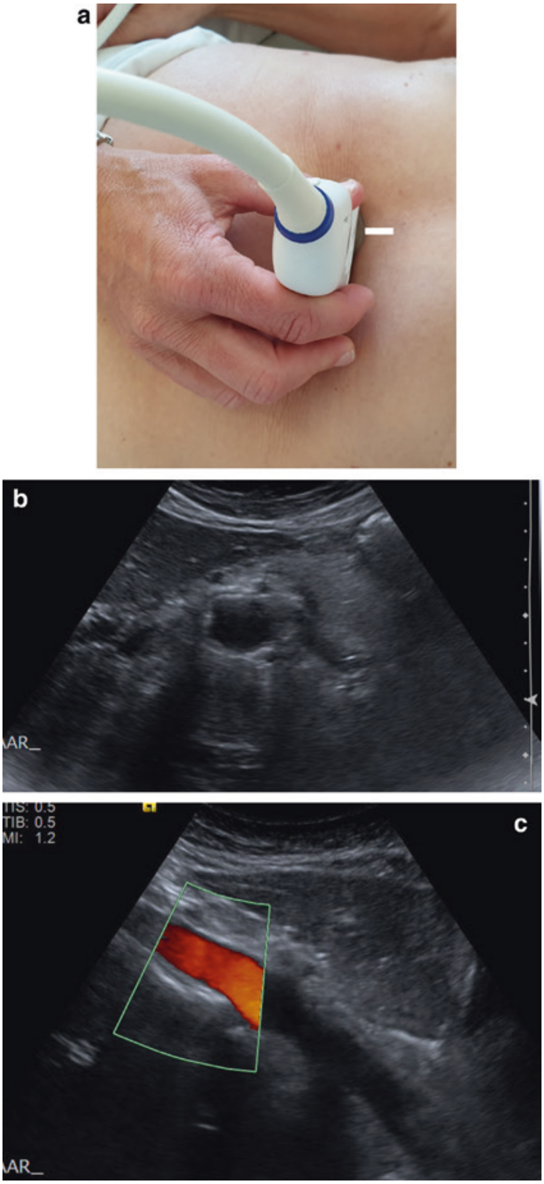


Fig. 20 Examination of **the abdominal aorta**: **a** recommended transducer position, **b** ultrasonographic image, transverse view **c** ultrasonographic image, longitudinal view

the common iliac arteries should be identified. The descending aorta is not satisfactorily visualized by transthoracic ultrasonographic examination.

D. **Pitfalls**

See Fig. 21, 22, and 23.

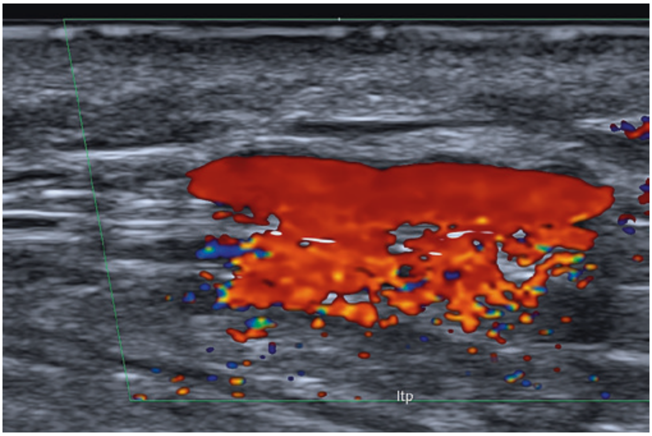


Fig. 21 Blooming: Excessive color which covers the vessel wall

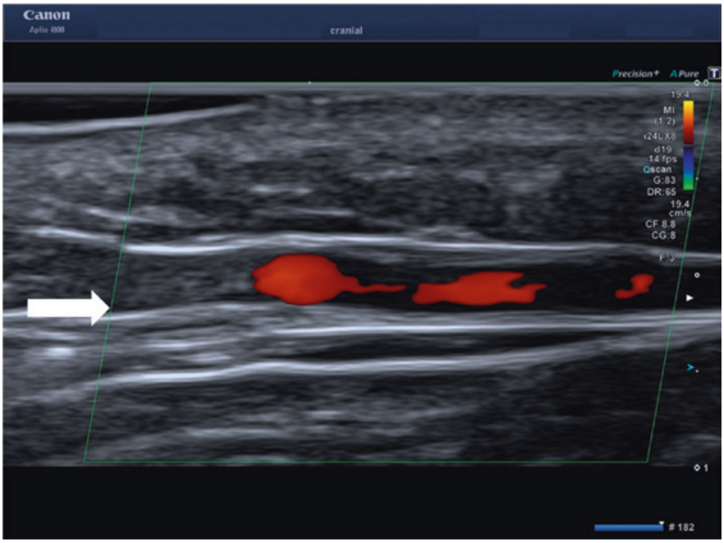


Fig. 22 Pseudohalo. Anechoic halo due to low color Doppler gain, high PRF or wrong angle of insonation (white arrow). The color doesn't fill the vessel lumen completely

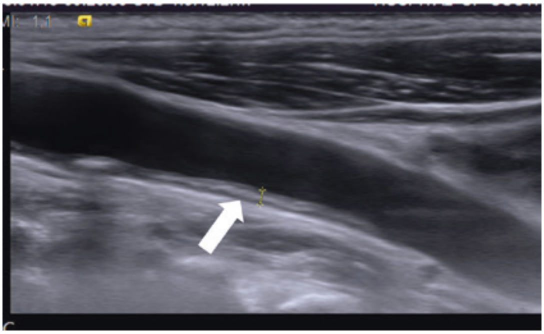


Fig. 23 Atherosclerosis in the carotid artery (white arrow)

Pathology

A. **Cranial Arteries**

See Figs. 24 and 25.

B. **Supraaortic Arteries**

See Fig. 26, 27, 28, 29, 30, and 31.

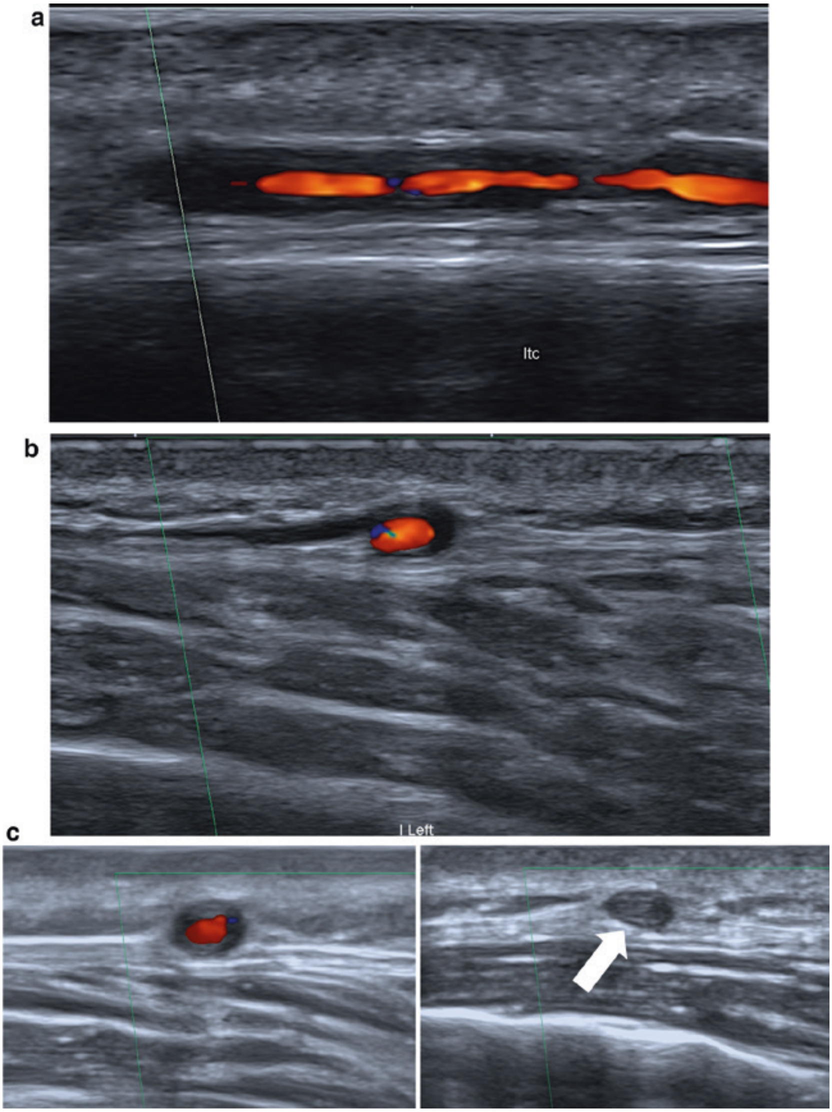


Fig. 24 Increased IMT in the common temporal artery in new-onset GCA. **a** Longitudinal, **b** transverse view **c** positive compression sign (white arrow)

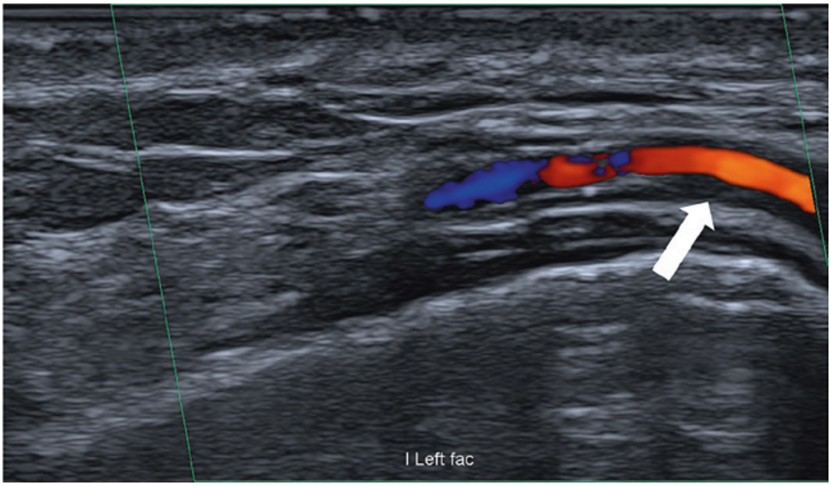


Fig. 25 Increased IMT in the facial artery, longitudinal view (white arrow)

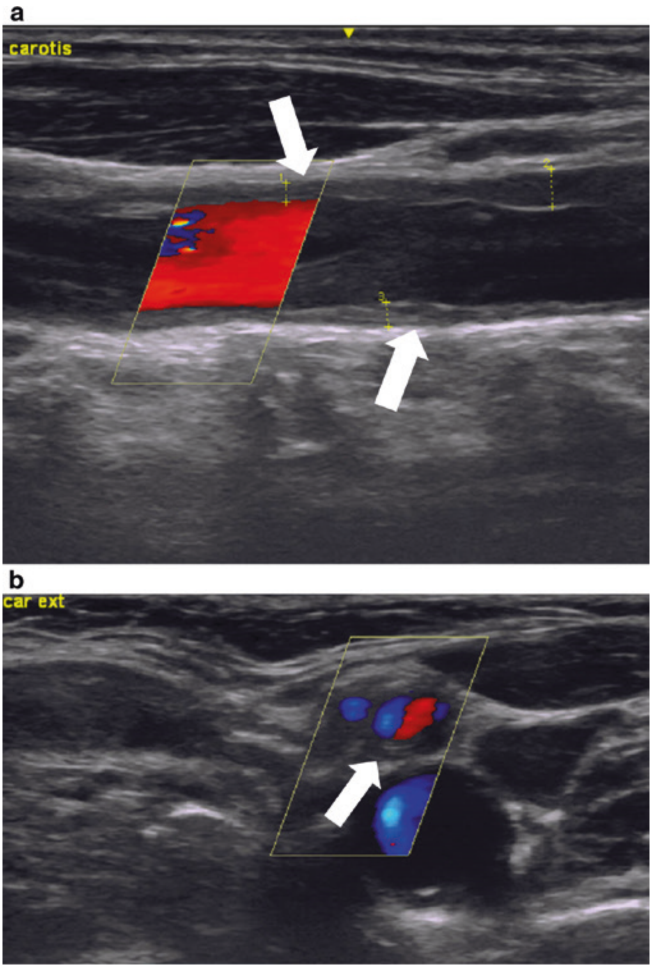


Fig. 26 Increased IMT in new-onset GCA: **a** in the common carotid artery **b** external carotid artery (white arrows)

Fig. 27 Increased IMT in the vertebral artery (white arrow)

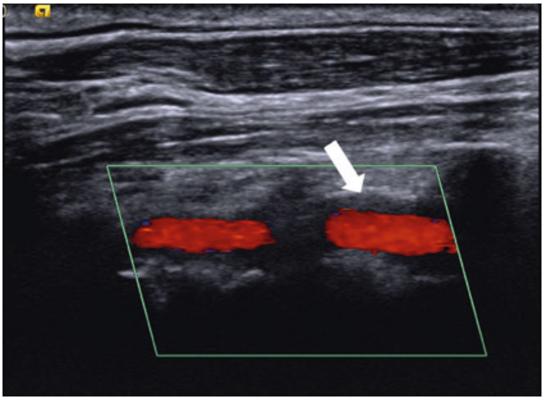


Fig. 28 Increased IMT in **a** the left subclavian artery, **b** the right subclavian artery

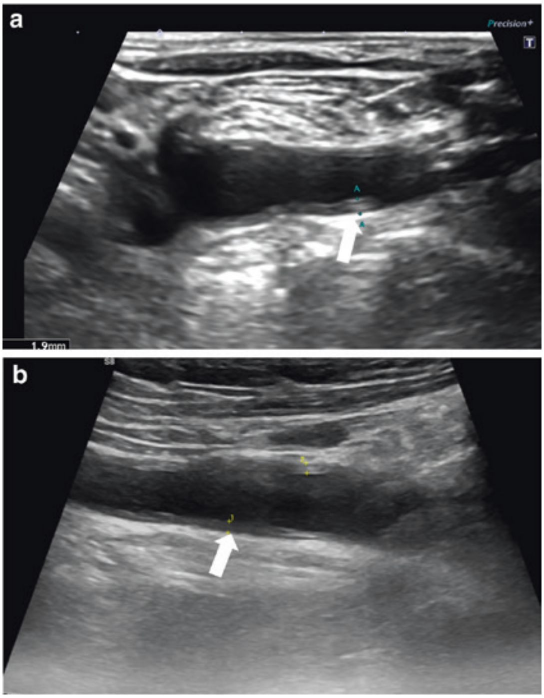
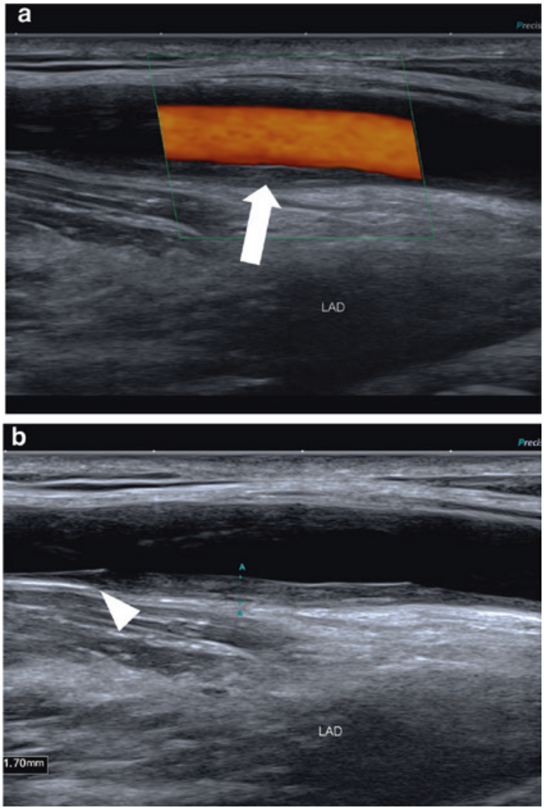


Fig. 29 Axillary arteritis (white arrow) in a GCA patient (axillary imaging approach) **a** distal axillary artery, **b** distal axillary artery, slope sign (white arrowhead at the distal part of the artery)



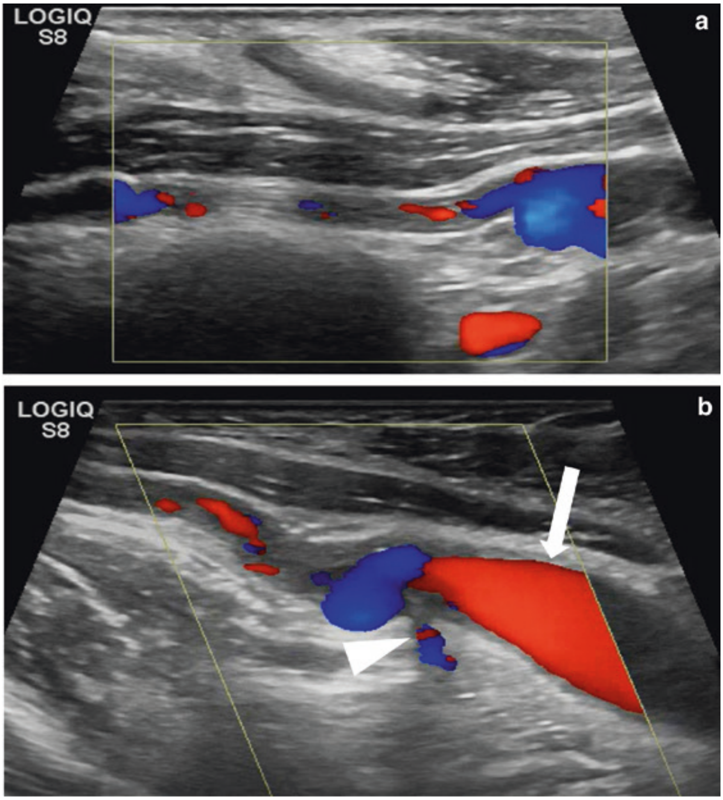
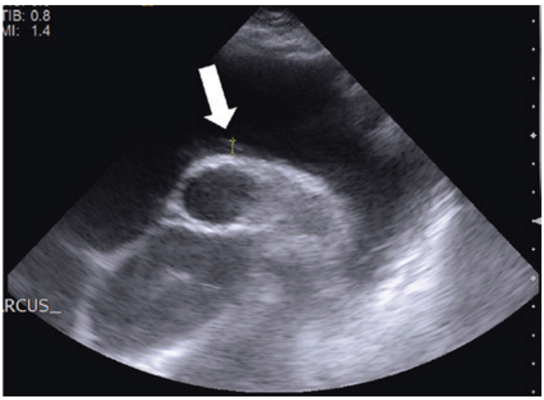


Fig. 30 Axillary artery occlusion in a patient with longstanding GCA (anteromedial imaging approach) **a** distal axillary artery, **b** proximal axillary artery. Note the area of aneurysm in the proximal axillary artery (white arrow) at the subscapular artery area (white arrowhead)

Fig. 31 Increased IMT measured at 5 mm in the aortic arch of a patient with new-onset GCA



Other Vasculitides

See Fig. 32.

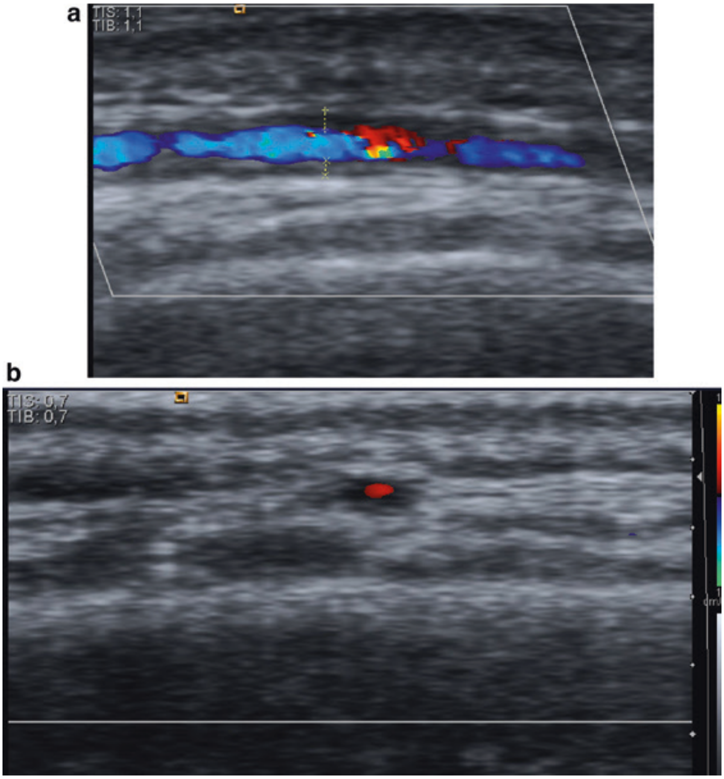


Fig. 32 Increased IMT in the parietal branch of the temporal artery in **a** granulomatosis with polyangiitis **b** polyarteritis nodosa

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