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Role of Temporal Artery Doppler and Halo Score in The Diagnosis, Prognosis and Monitoring of Giant Cell Arteritis in A Tertiary Care Centre-A Prospective Study

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Abstract:
Background: *Giant cell arteritis (GCA) is a common large vessel vasculitis of the elderly, often associated with sight loss.Despite the approval of biologic therapies, glucocorticoids (GC) continue to be the basis of therapy. In GCA, there are no biomarkers for disease severity, relapse probabilities, or damage. For suspected GCA, EULAR advises ultrasound (US) as the initial investigation. A non-compressible halo, the key Ultrasound discovery, is currently categorized as either positive or negative. In this study, we evaluate the hypothesis that the temporal and axillary arteries' halo signs, which are measured as a single composite Halo score (HS), may have diagnostic, prognostic, and monitoring value.*
Objective: *The aim of this study was to determine the diagnostic accuracy of temporal artery doppler and halo score in the diagnosis and prognosis of giant cell arteritis.*
Materials and Methods: *The study was conducted at Department of Radio-Diagnosis, Medical College Kolkata. A total of 27 patients with signs and symptoms of suspected giant cell arteritis were referred to department of Radio-diagnosis for colour duplex ultrasonography of temporal arteries. The study was conducted from 1st September 2021 to 31st August 2022. Conventional sonographic and doppler findings were documented and a composite halo score was calculated for each patient. The patients were treated based on the doppler findings and the halo score and were followed up during the study period.*
Results: *A total of 27 patients were included with a mean age of 52 years (range 33-64 years). Majority of the patients were male (66.7%). Bilateral halo sign in temporal arteries were seen in 21(77%) out of the 27 patients. Axillary artery involvement was seen in 11(40%) out of 27 patients. Sensitivity and specificity of halo score in diagnosing GCA was calculated to be 94.7 % and 75 % respectively. Majority of patients (85%) showed significant reduction in their halo scores on follow up scan. Follow-up study showed an 80% reduction of the vessel wall in temporal artery with treatment contrasted to the large vessels showing a reduction of 35%.*
Conclusion: *Our results show that Colour doppler ultrasonography is an inexpensive, non-invasive, reproducible, and easy-to-perform method that should precede temporal artery biopsy in all patients with suspected GCA.Biopsy is not necessary in a substantial proportion of patients in whom bilateral halo signs around the temporal arterial wall are found in Colour doppler studies.The halo score can be used as an efficient diagnostic, prognostic and disease monitoring tool for GCA. Ultrasound halo scoring allows to quantify the extent of vascular inflammation in GCA.*
Key word:*Temporal artery doppler; Giant cell arteritis; Ultrasound; Halo score: Halo sign*

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I. Introduction

Giant cell arteritis (GCA) is an autoimmune disease characterised by inflammation of large-sized and medium-sized arteries. Systemic inflammation, indicated by elevated C-reactive protein (CRP) levels, anaemia, and thrombocytosis, is frequently discovered through laboratory testing. A positive temporal artery biopsy (TAB) has been the gold standard for histological diagnosis of GCA. However, a biopsy is invasive, and it lacks sensitivity. According to EULAR recommendations, ultrasonography of the temporal and axillary arteries should be performed as soon as a patient is suspected of having GCA. The key ultrasonography result that pointsto GCA is a halo. A halo is a uniform, hypoechoic wall thickening of the artery that reflects oedema of the arterial wall brought on by inflammation. For GCA, ultrasound has a sensitivity of 77% and a specificity of

96%. The aorta and its big branches, such as the subclavian and axillary arteries, as well as branches of the external carotid arteries, such as the temporal arteries, are primarily involved in GCA. New headache, scalp discomfort, jaw claudication, diplopia, and amaurosis fugax are typical presenting symptoms. Significant morbidity and ischemia consequences from GCA, such as permanent blindness, are possible. Screening tests are essential since GCA symptoms are frequently vague and a wrong diagnosis might have disastrous consequences.

II. Review of Literature

When there is sufficient knowledge and equipment, the EULAR recommendations for imaging in large vessel vasculitis suggest using ultrasound of the temporal and/or axillary arteries as the initial imaging modality, especially in patients with suspected mainly cranial GCA. Given current EULAR recommendations that advise various diagnostic approaches in individuals with low, intermediate, or high GCA probability, estimation of GCA probability has become crucial. The diagnosis of GCA may be made in patients with a high clinical suspicion of the condition and an initial positive imaging test, such as a US, without the need for additional testing (e.g., biopsy or further imaging). The diagnosis of GCA can be deemed doubtful in patients with a low clinical probability and a negative imaging result, reassuring the patient. According to Aschwanden et al's assessment of 11 vascular areas for the presence of a halo, participation of the big systemic arteries is linked to greater weight loss. A low incidence of eye problems was associated with axillary artery involvement, according to Schmidt et al. In the latter trial, there was no correlation between the risk of eye problems and the proportion of haloed temporal artery (TA) segments. Halo thickness was not taken into account while determining the severity of the illness in either of these trials. The main finding on US in GCA patients is the halo sign: non-compressible hypoechoic wall swelling. When compared to the final clinical diagnosis of GCA, the most recent meta-analysis of prospective trials found that temporal artery ultrasound had a pooled sensitivity of 77% and a pooled specificity of 96%.

III. Materials And Methods

Patients:

The study was conducted at Department of Radio-Diagnosis, Medical College Kolkata. A total of 27 patients with signs and symptoms of suspected giant cell arteritis were referred to department of Radio-diagnosis for colour duplex ultrasonography of temporal arteries. The study was conducted from 1st September 2021 to 31st August 2022. Conventional sonographic and doppler findings were documented and a composite halo score was calculated for each patient. The patients were treated based on the doppler findings and the halo score and were followed up during the study period.

Study design:

In each patient, colour duplex ultrasonography was done using GE Logiq P9 ultrasound scanner equipped with L8-18i linear array small footprint transducer. Grey-scale frequency of 18 MHz and colour Doppler frequency of 9 MHz was used. For the Temporal artery, the focus was placed 5 mm below the skin. The pulse repetition rate ranged between 2 and 3 kHz.The colour box was set at an angle of at least 60°. The gain setting was adjusted to just fill the lumen. Patients were lying in a (semi-)recumbent position during the examination.The axillary arteries, the common superficial TA, and its parietal and frontal branches were all thoroughly and bilaterally examined in both the long and short planes. The biggest halo's thickness in each vascular region was calculated at its thickest point in the longitudinal plane.

Halo Score:

Halo score was assigned to each patient during the ultrasound examination.The halo sign at each branch of the common temporal, parietal and frontal arteries were scored 0–4 points, giving a maximum possible halo score (HS) score of 24 (Table 1). At the axillary arteries, the Intimal Medial Thickness (IMT) were scored 0–4 points on each side, allowing a maximum total score 8, which were multiplied by 3 (Fig. 2). A total halo score (THS) with maximum possible score of 48 was constructed by adding the scores of the temporal artery branches with the axillary artery score.

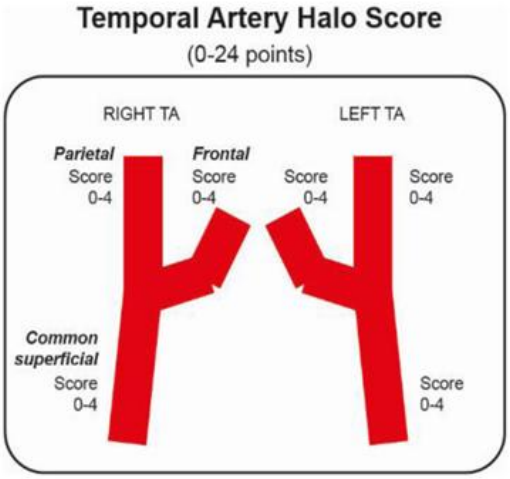

Fig 1. Diagram demonstrating the six temporal artery segments for calculating the temporal artery halo score.

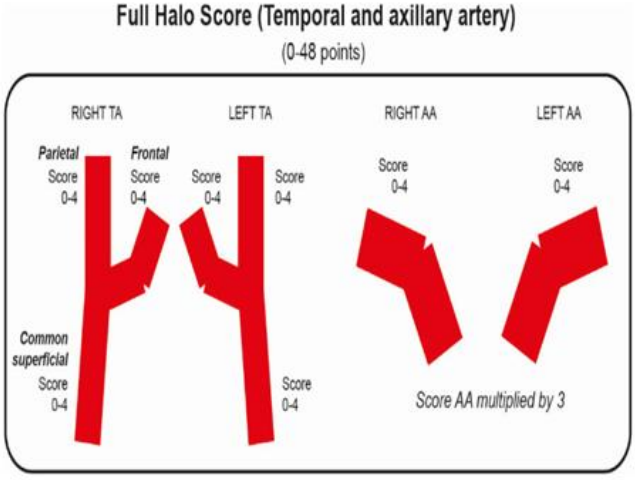

Fig 2.Diagram demonstrating the six temporal artery segments and two axillary arteries for calculating the total artery halo score.

Halo Grading	Common superficial TA halo thickness (mm)	Parietal TA halo thickness (mm)	Frontal TA halo thickness (mm)	Axillary artery halo thickness (mm)
Grade 0	0.3 or less	0.2 or less	0.1 or less	0.5 or less
Grade 1	0.4	0.3	0.2	0.6
Grade 2	0.5	0.4	0.3	0.7–0.8
Grade 3	0.6–0.7	0.5	0.4	0.9–1.5
Grade 4	0.8 or more	0.6 or more	0.5 or more	1.6 or more

Table 1: Halo score grading

Final diagnosis of GCA was done based on revised classification criteria as proposed recently (Dejaco et al. Rheumatology 2016) in the modified GiACTA criteria detailed below:
All of the following criteria has to be met:

- ESR > 30 mm/hr. or CRP > 10 mg/L.
- Unequivocal cranial symptoms of GCA (i.e., new-onset localised headache, scalp or temporal artery tenderness, ischemia-related vision loss, or otherwise unexplained mouth or jaw pain upon mastication) or symptoms of polymyalgia rheumatica (PMR), defined as shoulder and/or hip girdle pain associated with inflammatory morning stiffness.
- Evidence of GCA by imaging.

Inclusion Criteria:
1. Patients with unequivocal symptoms of GCA (i.e., new-onset localised headache, scalp or temporal artery tenderness, ischemia-related vision loss, or otherwise unexplained mouth or jaw pain upon mastication).
2. Patients must be more than 18 years of age.
3. Patients with informed consent for examination.

Exclusion Criteria:
1. Patients must not have a previous diagnosis of GCA.
2. Patients must not be under 18 years.
3. Inability to give informed consent.

Ethical Consideration:
The study was conducted after getting approval from Institutional Ethics Committee and other authority. Informed consent was taken from all participants.

IV. Results

This study included 27 patients with symptoms of GCA. Their ages ranged from 33 years to 64 years with a mean age of 52 years (table 2 and figure 3).

Age(years)	No.	%
30-<40	3	11.1
40-<50	9	33.3
50-<60	12	44.4
>=60	3	11.1
Min-Max	33-64	
Mean+- SD	52 +-10.7	
Median	51	

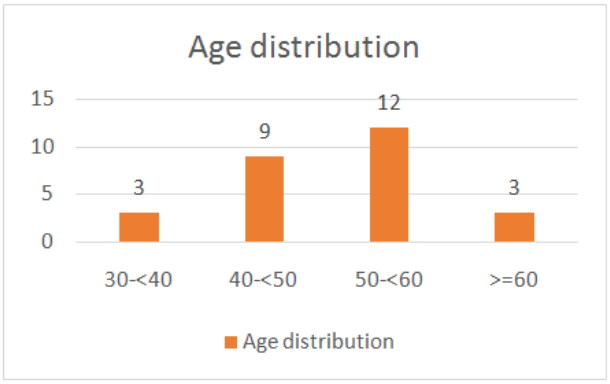

Table 2: Distribution of the patients according to age.Fig 3: Distribution of patients according to age.

Among the study population, 18 were male, while only 9 were female.

Sex	No.	%
Male	18	66.6
Female	9	33.3

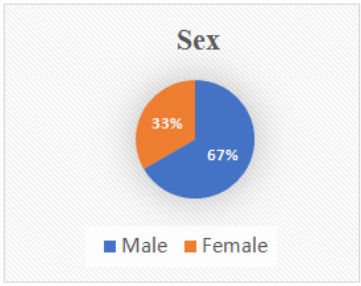

Table 3 and Fig 4: Distribution of the patients according to sex.

All patients were evaluated by colour duplex ultrasonography of bilateral temporal arteries and axillary arteries and were specifically examined for halo sign and a composite halo score was calculated for each of them.

Halo score	No. of patients
0-10	7
11-20	15
21-30	3
31-40	2
>40	0

Table 4: Distribution of patients according to halo score.

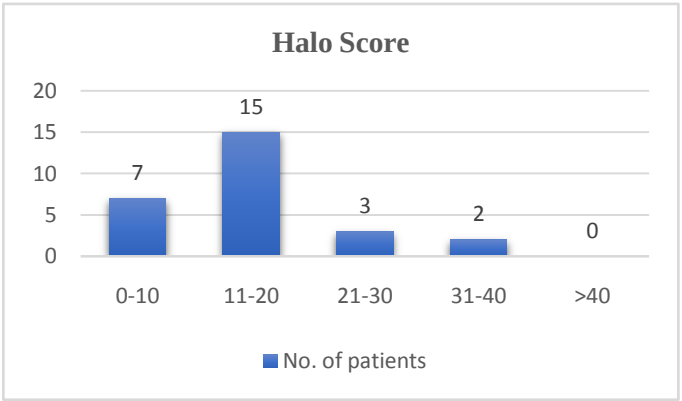

Fig 5: Distribution of patients according to halo score.

Bilateral halo sign in temporal arteries were seen in 21(77%) out of the 27 patients. Axillary artery involvement was seen in 11(40%) out of 27 patients.
All patients were further evaluated with inflammatory markers (ESR &CRP) and were finally diagnosed as GCA or not based on modified GiACTA criteria as mentioned above. Cut off Halo score of 10 was established for diagnosis of GCA.

Halo Score	Final Diagnosis		Total
	GCA	Not GCA	
<10	1	6	7
>=10	18	2	20
Total	19	8	27

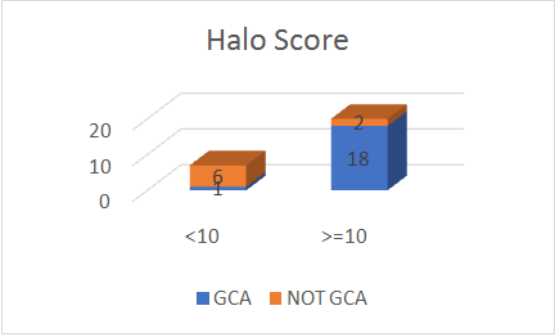

Table 5: Relationship between halo score and final diagnosis

Fig 6: Relationship between halo score and final diagnosis.

Sensitivity and specificity of halo score in diagnosing GCA was calculated to be 94.7 % and 75 % respectively. Positive and Negative predictive value was 90 % and 85% respectively, while accuracy was calculated to be 89%.
19 patients with diagnosed GCA were treated with long course steroids and other anti-inflammatory drugs and were followed up at 6-month interval with repeat temporal artery doppler and inflammatory markers. 2 patients were lost to follow-up. Halo score was calculated for 17 patients on follow up after treatment.

Halo score	No. of patients
0-10	6
11-20	9
21-30	2
31-40	0
>40	0

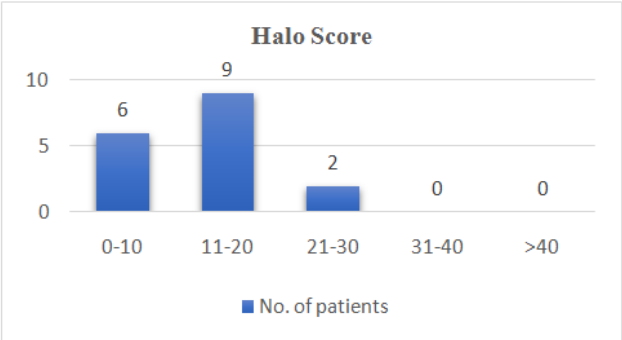

Table 6: Distribution of followed up patients according to halo score.

Fig 7: Distribution of followed up patients according to halo score.

Majority ofpatients (85%) showed significant reduction in their halo scores on follow up scan. Follow-up study showed an 80% reduction of the vessel wall in temporal artery with treatment contrasted to the large vessels showing a reduction of 35%.

Imaging

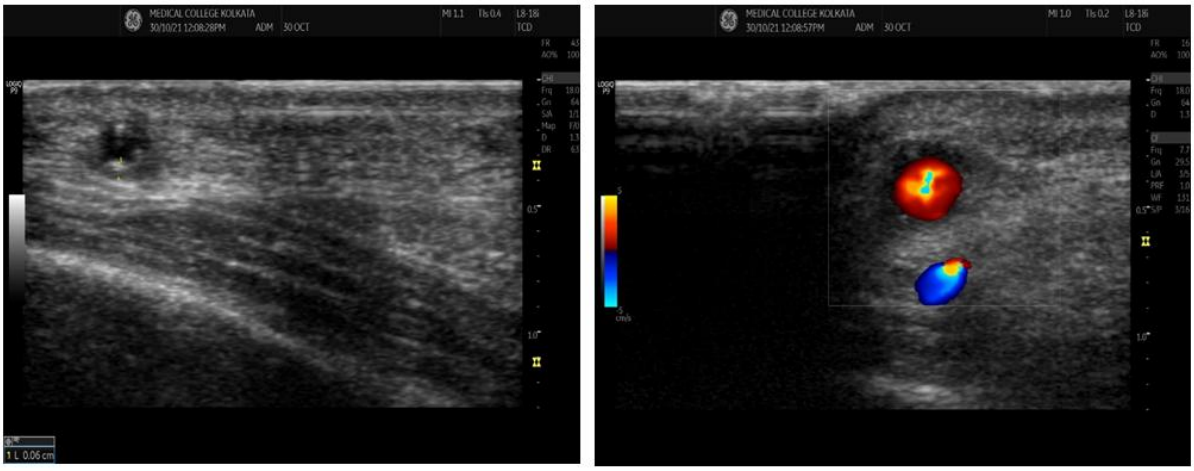

Fig 8 and 9: A 53-year-old male presented with severe throbbing headache more on right side. B mode showed increased diameter of the common superficial temporal artery.Colour doppler shows hypoechoic wall thickening characteristic of halo sign.Halo score was calculated to be 21 in this patient.

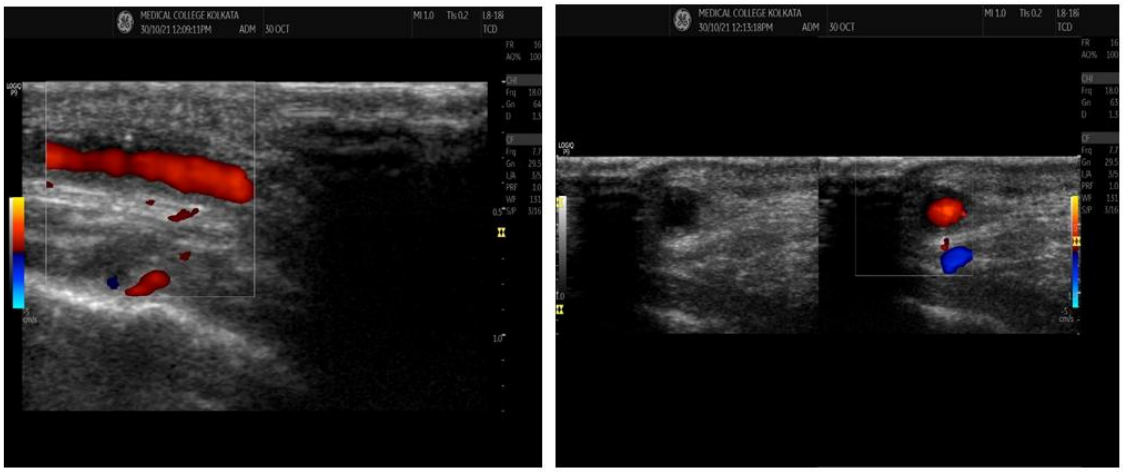

Fig 10 and 11: A 58-year-old female presented with left temporal headache and polyarthralgia. Temporal artery doppler showed typical pattern of circumferential hypoechoic wall thickening involving the painful superficial temporal artery. B mode and colour doppler showed edematous wall swelling of parietal part of temporal artery. Halo score was calculated to be 17 in this patient.

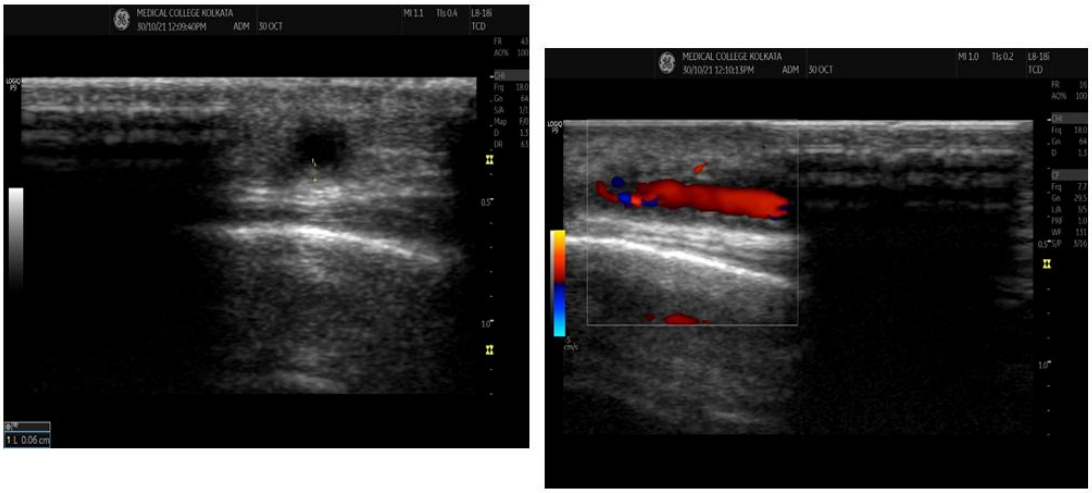

Fig 12 and 13: A 55-year-old male presented with severe headache, dimness of vision and jaw claudication. On ultrasound, the frontal part of temporal artery showed a hypoechoic wall thickening with a diameter of up to 0.6 mm around the perfused lumen. Halo score was calculated to be 31 in this patient.

V. Discussion

US is a non-invasive method that is safe, simple to perform, and repeatable in a clinic setting without subjecting the patient or the sonographer to radiation. BSR strongly advises US or TAB as a confirming test in suspected GCA, while EULAR recommends US as a first-choice imaging investigation.Halo is a dark hypoechoic area around the vessel lumen representing the vessel wall inflammation.

In this study of 27 suspected GCA patients, majority of the patients were in the age group of 50-60 years (44.4%) and were male (67%). Cut off Halo score of 10 was established for diagnosis of GCA. All patients were further evaluated with inflammatory markers (ESR &CRP) and were finally diagnosed as GCA or not based on modified GiACTA criteria.Bilateral halo sign in temporal arteries were seen in 21(77%) out of the 27 patients. Axillary artery involvement was seen in 11(40%) out of 27 patients. Sensitivity and specificity of halo score in diagnosing GCA was calculated to be 94.7 % and 75 % respectively. 19 patients with diagnosed GCA were treated with steroids and other anti-inflammatory drugs and were followed up at 6-month interval with repeat temporal artery doppler and inflammatory markers. 2 patients were lost to follow-up. Halo score was calculated for 17 patients on follow up after treatment. Majority of patients (85%) showed significant reduction in their halo scores on follow up scan. Follow-up study showed an 80% reduction of the vessel wall in temporal artery with treatment contrasted to the large vessels showing a reduction of 35%.

VI. Conclusion

Our results show that Colour doppler ultrasonography is an inexpensive, non-invasive, reproducible, and easy-to-perform method that should precede temporal artery biopsy in all patients with suspected GCA.Biopsy is not necessary in a substantial proportion of patients in whom bilateral halo signs around the temporal arterial wall are found in Colour doppler studies.The halo score can be used as an efficient diagnostic, prognostic and disease monitoring tool for GCA. Ultrasound halo scoring allows to quantify the extent of vascular inflammation in GCA. It is necessary and timely to identify prognostic variables in GCA. Recent BSR recommendations advise starting a high dose of steroids right away in individuals who have a high likelihood of having GCA.

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