Aberrant Right Subclavian Artery as Incidental Finding and Associated to Dysphagia. A series of Cases and Review of the Literature

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Abstract

The Aberrant right subclavian artery (ARSA), also known as the "arteria lusoria," is a distinctive congenital anomaly of the aortic arch. Its estimated prevalence in the general population ranges from 0.16% to 4.4%, with a higher incidence in women. Many people remain asymptomatic throughout their lives, and the anomaly is usually discovered incidentally during imaging studies. The symptoms typically present as non-progressive mechanical dysphagia, primarily with solid foods, and usually appear around the fifth decade of life. Computed tomography angiography (CTA) is an invaluable tool for identifying the anomalous origin, its retroesophageal course, and the degree of esophageal compression. In addition to anatomical imaging studies, functional esophageal studies are gaining recognition for their role in evaluating dysphagia and distinguishing between true pathological compression and incidental findings. We present a series of four clinical cases of an anomalous right subclavian artery, either alone or associated with dysphagia, incidentally diagnosed in adult patients by CT. Our objective is to perform a thorough literature review to thoroughly analyze the implications and significance of this vascular anomaly, with a focus on its relevance in cases of dysphagia or dyspnea and its application to patients scheduled for surgical procedures in the chest and neck regions.

Keywords: Arteria lusoria; Computed Tomography; Dysphagia; Incidental; Surgical Implications.

Introduction

Typically, the right subclavian artery originates from the brachiocephalic trunk. However, in the case of the lusory artery, it arises directly from the descending aorta, distal to the left subclavian artery. It follows a retroesophageal course until it reaches the right arm, where it can compress the posterior wall of the esophagus [4,5]. The prevalence of the lusoria artery in the general population is estimated to range from 0.16% to 4.4%. [9] Despite its relative frequency, between 60% and 80% of individuals with ARSA remain asymptomatic, and the anomaly is usually discovered incidentally during imaging studies. [4] Among the mechanisms explaining its symptomatology are

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increased vascular wall stiffness, aortic elongation, or the development of atherosclerotic disease, which can lead to increased extrinsic pressure on the esophagus. Additionally, the decline in the esophageal wall's flexibility and elasticity with age can render it more vulnerable to compression by the anomalous artery. [1,2,3]

Computed tomography angiography (CTA) is a useful diagnostic tool that allows for precise identification of the anomalous origin, its retroesophageal course, and the degree of esophageal compression. [6] CTA can also effectively detect associated complications, such as aneurysmal dilation or Kommerell's diverticulum, and even thrombosis, which can exacerbate esophageal obstruction. [2,12,13] The management of these cases is primarily dictated by the severity of symptoms and the presence of complications.[3] In cases of mild symptoms, medical management, such as proton pump inhibitors (PPIs), prokinetic agents, and dietary modifications, provides temporary relief. [5,6,7] Surgical intervention is considered for patients with intractable dysphagia, significant esophageal compression, or associated complications like aneurysmal dilation or thrombosis of the ARSA. [19]

Case Reports

Case 1

A 50-year-old female patient was referred to the ENT department of the external hospital with a left parotid mass that was suspected to be a Warthin tumor. A CT scan reveals an expansive mass in the superficial lobe of the parotid gland with maximum dimensions of approximately 24 × 32 × 20 mm in its transverse, anteroposterior, and cephalocaudal diameters. The mass demonstrates heterogeneous enhancement following intravenous contrast adminis-tration, a finding consistent with the clinical history. Incidentally, a tortuous retroesophageal course of the right subclavian artery, consistent with the Lusoria artery, was observed as an incidental finding.

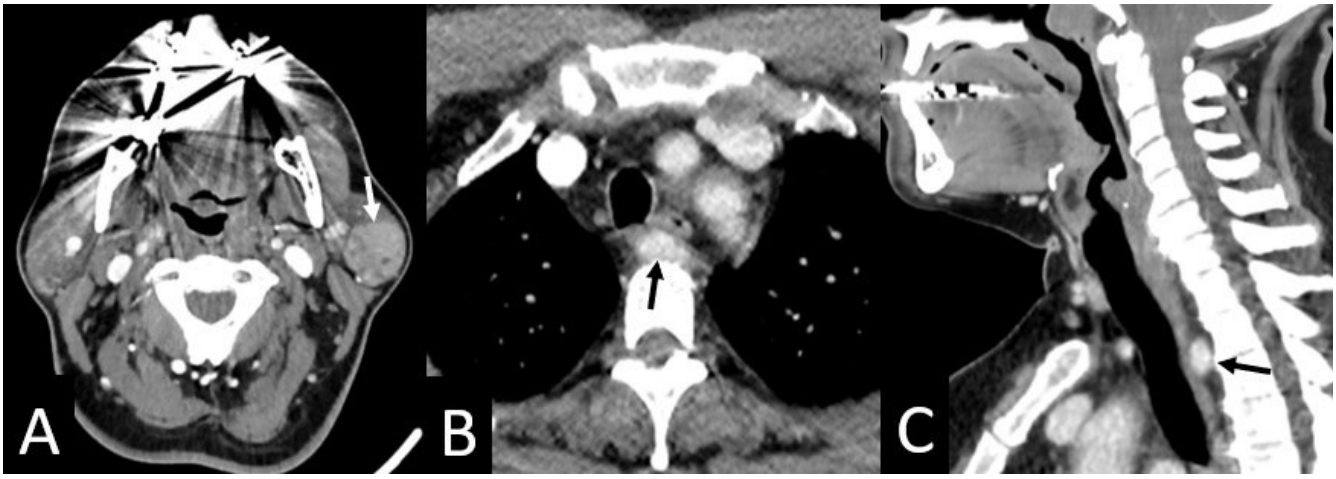


Fig. 1: Contrast-Enhanced CT Neck. A: soft-tissues mass in the left parotid gland (white arrow). B, C: the axial and sagittal views demonstrate an aberrant right subclavian artery (black arrows).

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Case 2

A 39-year-old female presented to the emergency department of an external hospital with suspected pharyngeal foreign body, with no other relevant medical history. A hyperattenuating intraluminal foreign body was identified on a CT scan at the level of the cervical esophagus in relation to the vertebral bodies from C7 to T2. The dimensions of the foreign body were determined to be approximately 27 × 2.3 × 30 mm in its transverse, anterior-posterior, and cephalocaudal diameters. It was found to be in close proximity to an aberrant right subclavian artery in a retroesophageal position as an incidental finding.

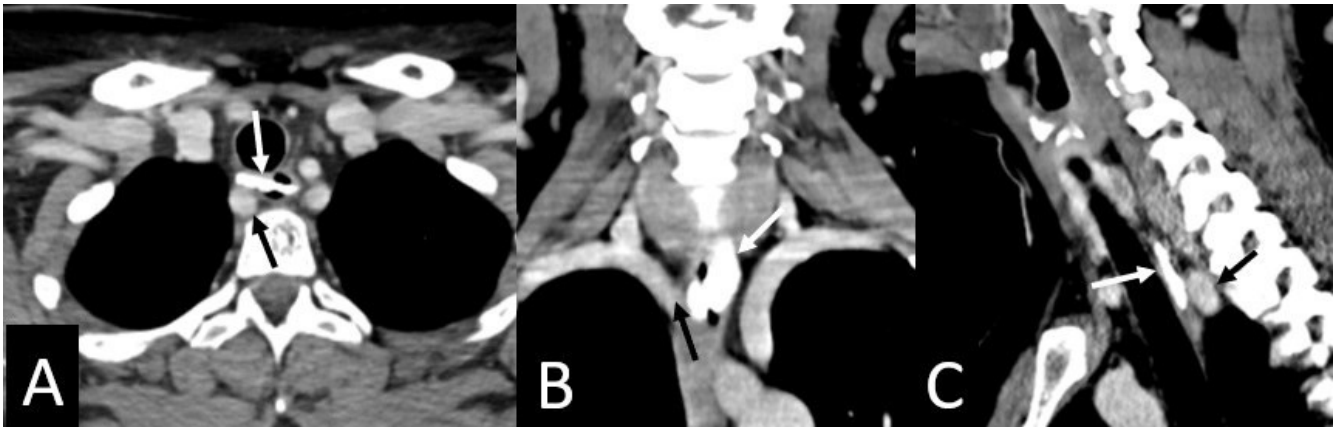


Fig. 2: Contrast-Enhanced CT Neck. hyperattenuating intraluminal foreign body in the cervical esophagus (white arrow) and its direct continuity with the anomalous right subclavian artery (black arrows). A, axial view; B, coronal view; and C, sagittal view.

Case 3

A 51-year-old female was admitted to a hospital emergency department with a clinical diagnosis of complicated bilateral acute otitis media, with suspected otomastoiditis, and a retropharyngeal abscess under investigation. A CT scan revealed the right subclavian artery, which was located immediately behind the trachea, resulting in extrinsic compression of the cervical esophagus. A thorough examination of the patient's CT temporal bone revealed no abnormalities.

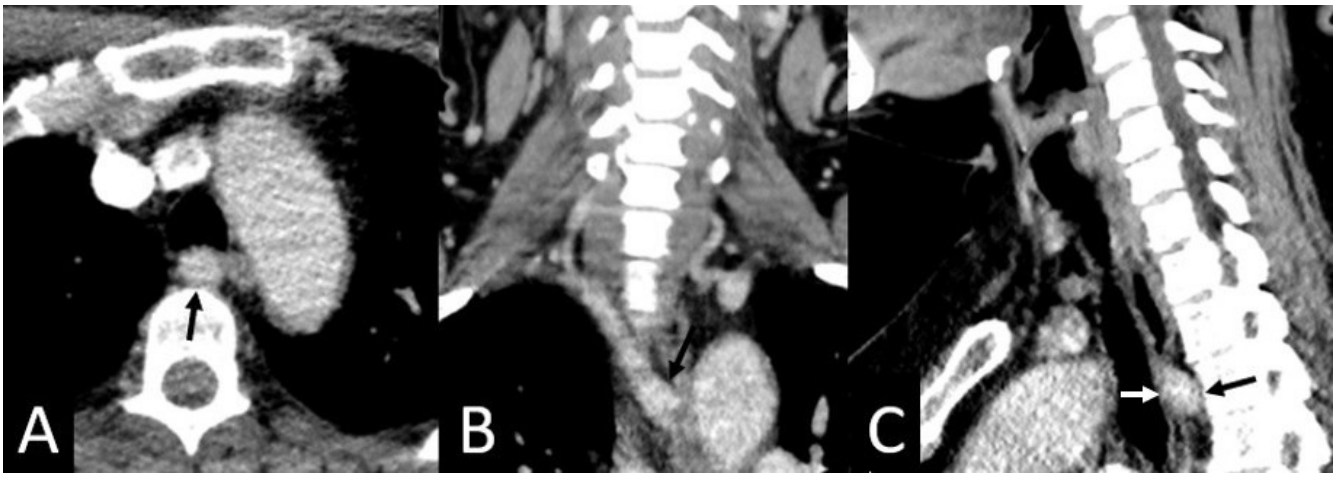


Fig. 3: Contrast-Enhanced CT Neck demonstrated an anomalous right subclavian artery (black arrows) with extrinsic compression of the cervical esophagus (white arrows). A, axial view; B, coronal view; and C, sagittal view.

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Case 4

An 82-year-old female patient was referred to the otolaryngology department of an external hospital for evaluation of symptoms related to dysphagia. The patient had no other significant medical history. A CT scan revealed an aberrant retrotracheal course of the right subclavian artery, which caused a rightward displacement and extrinsic compression of the cervical esophagus.

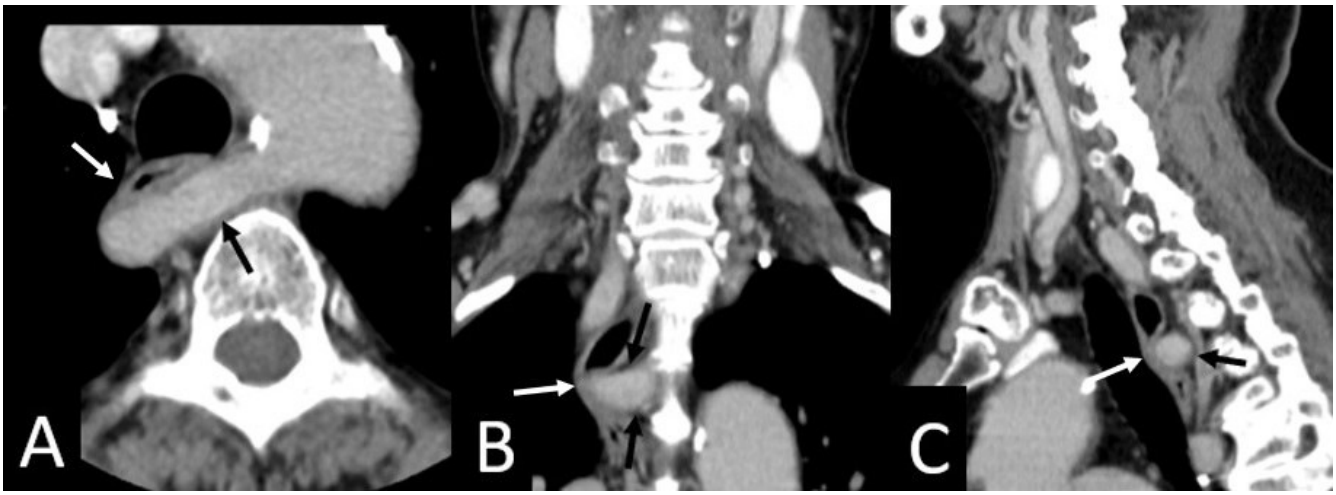


Fig. 4: Contrast-Enhanced CT Neck Contrast-enhanced CT of the neck showed an anomalous right subclavian artery (black arrows), resulting in rightward displacement and extrinsic compression of the cervical esophagus (white arrows). A, axial view; B, coronal view; and C, sagittal view.

Discussion

Dysphagia, defined as difficulty in swallowing, is a symptom that significantly impairs quality of life and can arise from a myriad of etiologies, encompassing neurological, functional, and structural disorders of the pharynx and esophagus. [1,2]

While common causes are frequently identified through standard diagnostic pathways, vascular anomalies represent a rare yet critical group of extrinsic esophageal compressions that can pose significant diagnostic challenges. [1,2] Among these, an aberrant right subclavian artery (ARSA), often referred to as arteria lusoria, stands out as the most prevalent congenital anomaly of the aortic arch. When this anomalous vessel compresses the esophagus, it results in a specific form of dysphagia known as dysphagia lusoria. [3] Recognizing this uncommon anatomical variant is paramount for accurate diagnosis and appropriate management, which can substantially improve patient nutrition and overall well-being. [1,2,3]

Arteria lusoria, is a distinct anatomical variant characterized by the anomalous origin of the right subclavian artery. Typically, the right subclavian artery originates from the brachiocephalic trunk; however, in arteria lusoria, it arises directly from the descending aorta, distal to the left subclavian artery. [4] This malformation is typically attributed to the persistence of the right dorsal aorta and distal right fourth aortic arch,

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coupled with the regression of the proximal right fourth aortic arch. [5] This aberrant artery then typically follows a retroesophageal course to reach the right arm, potentially compressing the posterior wall of the esophagus. [5] The term "dysphagia lusoria" was coined in 1794 by David Bayford, who described a case of fatal deglutition obstruction attributed to this anomalous artery. [6] Although ARSA is the most common anomalous artery implicated in esophageal compression, other aortic arch variants, such as a right-sided aortic arch with an aberrant left subclavian artery, can also fall under the umbrella of dysphagia lusoria. [7]

Several associated vascular anomalies can further complicate the presentation of arteria lusoria. A proximal dilation of the aberrant artery, known as Kommerell's diverticulum, which is a remnant of the right fourth aortic arch, is concomitantly observed in approximately 20-60% of ARSA cases and can exacerbate esophageal compression. [8] Furthermore, an aneurysm of the ARSA, which occurs in about 13% of cases, can contribute to severe symptoms and often accounts for a delayed presentation of dysphagia. [9] Other anatomical variations, such as a common stem of the right and left carotid artery (bicarotid trunk), are found in approximately 19% of individuals with ARSA and may influence the clinical picture. [9]

The lusoria artery has an estimated prevalence in the general population ranging from 0.16% to 4.4% according to cadaveric and angiographic studies. [9] Despite its relatively common occurrence as an anatomical variant, the majority of individuals (60– 80%) with ARSA remain asymptomatic throughout their lives, with the anomaly often discovered incidentally during imaging for other conditions or upon autopsy. [4]

Symptomatic presentation typically manifests as nonprogressive mechanical dysphagia, primarily to solids, often occurring around the fifth decade of life. [2,3,4] A female predominance has been reported among symptomatic patients with dysphagia lusoria. [3] Less common symptoms include retrosternal chest pain, dyspnea, regurgitation, and coughing. [9] Some patients report symptomatic relief in the supine position, which can help differentiate it from gastroesophageal reflux disease (GERD). [11]

In children, the clinical presentation of dysphagia lusoria often differs from that observed in adults, with respiratory symptoms being more prevalent due to the close anatomical relationship between the esophagus and trachea in the pediatric mediastinum. [2,9,10,11,12] Infants and young children may present with recurrent respiratory infections, chronic cough, stridor, wheezing, and dyspnea, indicative of tracheal compression by the aberrant vessel or associated vascular rings. [2,9,10,11,12]

The mechanisms underlying the development of symptoms in only 20–40% of individuals with ARSA are multifactorial. [2] One prominent theory suggests that coexisting vascular anomalies, such as a bicarotid trunk or Kommerell's diverticulum, can further exacerbate the compression. [10] Age-related vascular changes are also implicated, where increasing rigidity of the vessel wall, elongation of the aorta, or development of atherosclerotic disease can lead to enhanced extrinsic pressure on the esophagus, thereby triggering symptoms in older adults. [1] Similarly, a decrease in the

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pliability and elasticity of the esophageal wall with advancing age may render it more susceptible to compression by the aberrant artery. [1,2,3]

Furthermore, there is a higher observed prevalence of ARSA in pediatric populations with specific chromosomal abnormalities, including Down syndrome, William's syndrome, and DiGeorge syndrome, as well as in those with congenital heart disease. [9] Consequently, a heightened clinical suspicion for dysphagia lusoria is warranted when evaluating patients with these comorbidities who present with dysphagia or other related symptoms, even in younger age groups. Early recognition in these vulnerable populations is crucial for timely intervention and prevention of severe complications. [9]

Initial diagnostic evaluation often includes non-invasive imaging modalities such as a barium esophagram, which can reveal a characteristic posterior indentation of the esophageal wall caused by the aberrant artery. [1] However, definitive diagnosis and detailed anatomical assessment rely on advanced cross-sectional imaging. [5,6,7,8,9]

Computed Tomography Angiography (CTA) of the chest is particularly useful for identifying the aberrant origin, its retroesophageal course, and the degree of esophageal compression. [6] CTA can also effectively detect associated complications such as aneurysmal dilation of the aberrant artery or Kommerell's diverticulum, and even thrombosis, which can exacerbate esophageal obstruction. [2,12,13] Magnetic Resonance Angiography (MRA) offers similar utility, providing detailed visualization of the retroesophageal course of the anomalous artery and identifying potential vascular mediastinum shadow, which can be an initial suggestive finding. [15] Cardiac MRI can further demonstrate associated anomalies, such as the ligamentum arteriosum in cases of right aortic arch with aberrant left subclavian artery. [13,14,15,16] Endoscopic ultrasound (EUS) has also been described for direct visualization of the arteria lusoria and assessment of its compressive effect on the esophagus. [17]

Beyond anatomical imaging, functional esophageal studies are increasingly recognized for their role in evaluating dysphagia lusoria and differentiating true pathological compression from incidental findings. [2] High-resolution manometry (HRM) and impedance manometry are valuable tools for assessing esophageal motility patterns, as some patients with dysphagia lusoria may exhibit dysmotility. [7,8,9] These tests help determine if the esophageal compression contributes to, or is the sole cause of, the dysphagia, especially when co-existing conditions like GERD are present. [17]

Esophagogastroduodenoscopy (EGD) may also be performed to rule out other concomitant structural or inflammatory causes of dysphagia that might mimic or complicate the presentation of dysphagia lusoria. [17] For cases involving rare vascular anomalies, such as an anomalous vertebral artery originating from the aortic arch, comprehensive diagnostic imaging remains vital for precise identification of the causative vessel. [15,16,17] The collective use of these diagnostic examinations provides a thorough understanding of the vascular anomaly, its impact on esophageal function,

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and guides appropriate management, ranging from dietary modifications to complex surgical interventions for severe or refractory symptoms. [18]

Management strategies for dysphagia lusoria are primarily dictated by the severity of symptoms and the presence of complications. [3] For patients presenting with mild symptoms, medical management often provides temporary relief. This conservative approach includes dietary modifications, along with pharmacological interventions such as proton pump inhibitors (PPIs) and prokinetic agents. [5,6,7] However, medical therapy alone may be insufficient for those with severe, persistent, or complicated dysphagia lusoria. [9]

Surgical intervention is generally considered for symptomatic patients, particularly those with intractable dysphagia, significant esophageal compression, or associated complications like aneurysmal dilation or thrombosis of the ARSA. [19] The primary objective of surgical correction is to relieve esophageal compression. Traditional surgical approaches often involve the division of the aberrant subclavian artery, which can effectively decompress the esophagus. [19] However, simple division without revascularization can lead to long-term hypotrophy or ischemia of the thoracic limb due to compromised blood flow. [20] To mitigate this risk, division is frequently followed by revascularization, where the divided aberrant subclavian artery is reimplanted into another vessel, such as the right carotid artery, via a supraclavicular approach. [21] This cervical approach allows for ligation of the artery near its root and subsequent connection to the right carotid artery. [20,21]

More complex cases, especially those involving Kommerell's diverticulum or aneurysmal dilation of the aortic arch or the proximal ARSA, may necessitate more extensive procedures. [22] Open surgical repair, hybrid endovascular approaches, and purely endovascular techniques are all considered based on patient anatomy and comorbidities. [23] Hybrid procedures combine open surgical debranching (e.g., creation of bypasses from the carotid artery to the subclavian artery) with subsequent endovascular stent-graft placement within the aorta to cover the origin of the aberrant vessel. [24] Endovascular repair alone, typically involving stent-graft deployment, is an option, especially for aneurysmal disease, but requires careful planning to ensure perfusion to the subclavian artery through either a bypass or a fenestrated/branched graft. [20] Minimally invasive approaches, such as staged robotic video-assisted thoracoscopic surgery (VATS), have also been described, offering potential benefits in recovery. [25] Outcomes of surgical repair are generally favorable, with many patients becoming symptom-free postoperatively, though intermittent dysphagia can persist in some cases. [25] The choice of surgical technique is highly individualized, considering factors such as the patient's overall health, the presence and morphology of Kommerell's diverticulum, and the extent of aortic involvement. [20,21,22,23,24,25]

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Conclusions

In summary, ARSA is a rare embryological abnormality. While it is usually asymptomatic, it can cause significant symptoms, such as dyspnea or dysphagia. It is essential to have a comprehensive understanding of anatomical and morphological variations through imaging to ensure accurate diagnosis and effective management. Medical therapy is suitable for mild symptoms, but surgical intervention, with various open, endovascular, or hybrid approaches tailored to the individual's specific anatomy and symptom severity, is often required for definitive relief and to prevent complications.

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Declarations

Consent for publication: The author clarifies that written informed consent was obtained and the anonymity of the patient was ensured. This study submitted to Swiss J. Rad. Nucl. Med. has been conducted in accordance with the Declaration of Helsinki and according to requirements of all applicable local and international standards. All authors contributed to the conception and design of the manuscript, participated in drafting and revising the content critically for important intellectual input, and approved the final version for publication. Each author agrees to be accountable for all aspects of the work, ensuring its accuracy and integrity.

Conflict of interest:

The authors declare that there were no conflicts of interest within the meaning of the

recommendations of the International Committee of Medical Journal Editors when the article was written. Journal Editors when the article was written.

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