International Multi-Institutional Experience with Presentation and Management of Aortic Arch Laterality in Aberrant Subclavian Artery and Kommerell's Diverticulum

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Background: Aberrant subclavian artery (ASA) with or without Kommerell's diverticulum (KD) is a rare anatomic aortic arch anomaly that can cause dysphagia and/or life-threatening rupture. The objective of this study is to compare outcomes of ASA/KD repair in patients with a left versus right aortic arch.

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Methods: Using the Vascular Low Frequency Disease Consortium methodology, a retrospective review was performed of patients ≥18 years old with surgical treatment of ASA/KD from 2000 to 2020 at 20 institutions.
Results: 288 patients with ASA with or without KD were identified; 222 left-sided aortic arch (LAA), and 66 right-sided aortic arch (RAA). Mean age at repair was younger in LAA 54 vs. 58 years ($P = 0.06$). Patients in RAA were more likely to undergo repair due to symptoms (72.7% vs. 55.9%, $P = 0.01$), and more likely to present with dysphagia (57.6% vs. 39.1%, $P < 0.01$). The hybrid open/endovascular approach was the most common repair type in both groups. Rates of intraoperative complications, death within 30 days, return to the operating room, symptom relief and endoleaks were not significantly different. For patients with symptom status follow-up data, in LAA, 61.7% had complete relief, 34.0% had partial relief and 4.3% had no change. In RAA, 60.7% had complete relief, 34.4% had partial relief and 4.9% had no change.
Conclusions: In patients with ASA/KD, RAA patients were less common than LAA, presented more frequently with dysphagia, had symptoms as an indication for intervention, and underwent treatment at a younger age. Open, endovascular and hybrid repair approaches appear equally effective, regardless of arch laterality.

INTRODUCTION

Aberrant subclavian artery (ASA) is a rare anatomic anomaly of the aortic arch, with an estimated prevalence of 0.8%−1%.[1−3] Patients with ASA may also have an associated Kommerell's diverticulum (KD), an aneurysmal dilation of the descending thoracic aorta at the origin of the ASA. Patients with ASA/KD most often have a left-sided aortic arch (LAA) with a right subclavian artery (RSA) that originates distal to the left subclavian artery. Less common is a right-sided aortic arch (RAA) with an aberrant left

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subclavian artery where the left common carotid originates first, followed by the right common carotid, right subclavian, then left subclavian. (Fig. 1).

In normal embryologic development, the right fourth arch becomes the left descending aorta and the right dorsal aorta fuses with the right seventh intersegmental artery to form the RSA.[4] In LAA with right-sided ASA, the primitive right arch abnormally involutes between the right subclavian and right common carotid, leaving the right subclavian as the most distal aortic branch. In RAA with left-sided ASA, the right fourth arch becomes the RAA instead of the left descending aortic arch, and the left-sided ASA develops due to the abnormal involution of the left fourth arch between the left subclavian and left common carotid.[5]

RAA occurs in only 0.01 to 0.1% of the general population and is more commonly associated with other congenital anomalies and embryological syndromes, such as tetralogy of Fallot and truncus arteriosus, and chromosomal abnormalities such as DiGeorge syndrome. The existing evidence comparing anatomic variations, presentation and surgical outcomes of ASA/KD is primarily in the form of case reports and series with a limited sample size.[6] While there have been several case reports and smaller studies of patients[7–12] looking specifically at surgical treatment of RAA, there are few studies and case series that include both LAA versus RAA for comparison.[13–17] The largest study population of operative treatment with ASA identified in our literature review is a single institution retrospective study of 65 open or endovascular repairs.[18] Given the limited data on the impact of arch laterality, this study was undertaken to evaluate the differences in presentation, surgical treatment, and outcomes between patients with LAA versus RAA who undergo repair of ASA/KD. We hypothesize that ASA/KD in RAA may present differently from LAA and may be more complex to treat surgically with potentially worse outcomes. Our international, multi-institutional cohort provides a unique opportunity to compare patients with LAA and RAA with ASA/KD with a robust study population and defined study protocol.

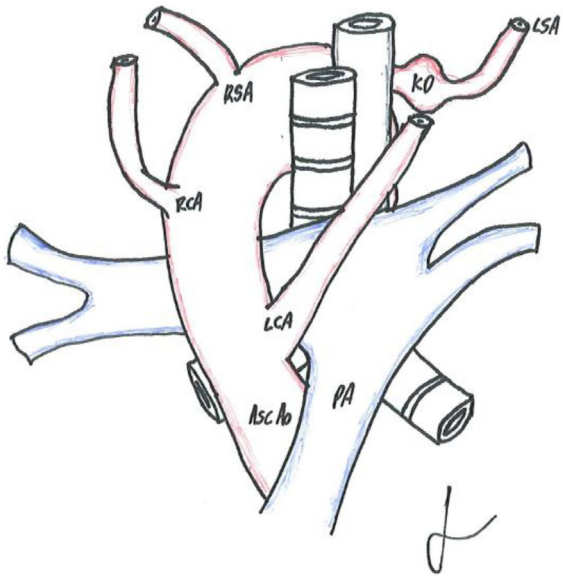

Fig. 1. Most Common Anatomy of RAA with ASA. AscAo, Ascending aorta; PA, pulmonary artery; LCA, left carotid artery; RCA, right carotid artery; KO, kommerell; LSA, left subclavian artery.

METHODS

Study Design

A retrospective review was performed using the Vascular Low Frequency Disease Consortium (VLFDC). The VLFDC is a collaborative effort among investigators from institutions across the world to study uncommon vascular conditions.[19] Each participating institution obtained local institutional review board (IRB) approval. After IRB approval, deidentified data were collected using a standardized data collection form. The study data were collected and managed using the Research Electronic Data Capture (REDCap®) tool hosted at the University of California, Los Angeles.[20] The data points and definitions were determined by the authors who developed the research project (Bath/D'Oria) in conjunction with the VLFDC team. The VLFDC investigators were responsible for the accuracy and completeness of data at their own institutions. Discrepancies were clarified with investigators and each investigator reviewed the associated abstract and manuscript prior to submission.

The inclusion criteria were patients ≥18 years of age who underwent open, endovascular, or hybrid surgical treatment of ASA, with or without KD, and with or without concomitant thoracic aortic disease, from January 1, 2000, to December 31, 2020. All cases were included regardless of initial presentation, involving asymptomatic, symptomatic, or ruptured aneurysms. All surgical approaches were included. Participating institutions identified eligible patients through ICD-9 and ICD-10 codes related to thoracic aortic aneurysm and anomalies, anomalous origin of subclavian arteries, and Current Procedural Terminology codes for open or endovascular repair, resection or replacement of

thoracic aorta, and carotid-subclavian bypass or subclavian-carotid transposition, institutional databases, or other record-keeping mechanisms (Appendix A).

Patient Data

Patient demographic data was recorded, including sex, age, race, and ethnicity (Appendix B). Comorbidities, prior surgical procedures, and preoperative patient presentation were obtained. ASA/KD transverse diameter was measured using preoperative computed tomography angiography, defined as the distance from the KD outer wall transversely to the opposite aortic wall. Surgical details were documented, including the urgency of the operation, surgical specialties involved and operative technique (open, hybrid, endovascular). Open technique was further classified as primary versus patch versus graft repair with ASA ligation versus carotid-subclavian bypass or subclavian-carotid transposition. Hybrid technique was further classified as carotid-subclavian bypass + thoracic endovascular aortic repair (TEVAR), carotid-subclavian bypass + TEVAR + ASA coil embolization, or subclavian-carotid transposition + TEVAR. Endovascular only repair was classified as TEVAR only or TEVAR with chimney/snorkel versus fenestrated/branched graft versus in-situ laser fenestration versus physician modified TEVAR graft. Anesthesia type, surgery duration and estimated blood loss were also recorded. Intraoperative and postoperative complications, both short-term and long-term were documented. Short-term complications included death, unexpected readmission, return to operating room, unplanned reintervention, endoleak or other acute event, within 30 days. Long-term data points collected included symptom relief within 1 year, ASA/KD diameter >6 months postoperatively, reintervention, death after 30 days, or endoleak.

Statistical Analysis

Continuous variables were described using mean and standard deviation or median and interquartile range, as appropriate. Categorical variables were compared using the Chi square or Fisher exact test, as appropriate. Continuous variables were compared using Student's *t*-test or Mann–Whitney *U*-test, as appropriate. Statistical significance was set at $P < 0.05$. Statistical analysis was performed using SAS 9.4 (SAS Institute Inc., Cary, NC).

RESULTS

Demographic and Baseline Clinical Presentation

The study cohort that met inclusion criteria consisted of 288 patients drawn from 20 institutions. Of these, 222 patients had a LAA, and 66 patients had a RAA (Table I). The mean age at repair was 57.9 vs 53.5 years in LAA and RAA patients, respectively ($P = 0.06$). There was no significant difference with respect to sex or identified race/ethnicity. With regards to baseline comorbidities, 7.2% of LAA versus 0% of RAA patients had congestive heart failure ($P = 0.02$). 12.8% of LAA versus 3.1% of RAA patients had chronic kidney disease ($P = 0.03$). There were no significant differences in the incidence of diabetes, hypertension, prior myocardial infarction, or concomitant arterial atherosclerotic or aneurysmal disease between groups (Table I). While not statistically significant, 16.1% of LAA and 9.1% of RAA patients had a prior coronary revascularization ($P = 0.16$); 17.6% of LAA and 10.6% of RAA patients had undergone prior aortic procedures ($P = 0.17$). In this dataset, LAA patients had a lower incidence of an associated KD compared to RAA, (66.0% vs. 85.7%, $P < 0.01$). Mean preoperative KD diameter was 38.7 mm in LAA and 49.0 mm in RAA patients respectively ($P = 0.33$). A greater proportion of RAA patients underwent repair of their ASA/KD for a symptomatic indication compared to those with LAA (72.7% vs. 55.9%, $P = 0.01$) with dysphagia being most common (RAA 57.6% vs. 39.1%, $P < 0.01$). Other symptoms that were recorded, including pain, dyspnea, dysphonia, and neurologic symptoms were not significantly different between the 2 groups.

Intraoperative and Postoperative Approach and Outcomes

The most common type of repair in both groups was the hybrid approach (Table II). Specifically, carotid-subclavian bypass with TEVAR with coil embolization was the most common repair among LAA patients (24.1%). Carotid-subclavian bypass and TEVAR without coil embolization was the most common approach in RAA (16.9%). A total of 25 patients underwent bilateral subclavian artery transposition or bypass, with a majority occurring among LAA patients (23 of 25, 10% of total vs. 2 (3%) of RAA patients). Moreover, 5 LAA patients (2%) underwent combined carotid-carotid and carotid-subclavian bypass or a total arch debranching procedure, compared to 2 (3%) RAA patients. Open repair was the second most common approach

Table I. Patient demographics by ASA aortic arch laterality, $N = 288$

Variable	*n* (%), left arch $N = 222$	*n* (%), right arch $N = 66$	*P* value
Median age at surgery	57.9	53.5	0.06
Sex			0.30
Male	115 (51.8)	39 (59.1)	
Female	107 (48.2)	27 (40.9)	
Race/ethnicity			0.16
Non-Hispanic White	179 (80.6)	53 (81.5)	
Black	23 (10.4)	5 (7.7)	
Hispanic	13 (5.9)	1 (1.5)	
Asian	3 (1.4)	2 (3.1)	
Other	4 (1.8)	4 (6.2)	
Comorbidities			
Diabetes	29 (13.1)	8 (12.3)	0.87
Hypertension	157 (71.4)	44 (66.7)	0.46
Congestive heart failure	16 (7.2)	0 (0.0)	0.02
Chronic kidney disease	28 (12.8)	2 (3.1)	0.03
Prior myocardial infarction <6 months of intervention	5 (2.5)	1 (1.5)	0.25
Prior myocardial infarction >6 months of intervention	31 (15.2)	5 (7.6)	0.25
Prior PCI/CABG	33 (16.1)	6 (9.1)	0.16
Prior aortic procedures	39 (17.6)	7 (10.6)	0.17
Concomitant arterial disease	72 (32.7)	19 (28.8)	0.55
KD present	132 (66.0)	54 (85.7)	0.003
Mean ASA/KD preoperative diameter, mm (95% CI)	38.7 (33.8–43.6)	49.0 (28.5–69.6)	0.33
Indication for surgery			
Size	80 (36.0)	28 (42.4)	0.35
Symptoms	124 (55.9)	48 (72.7)	0.01
Dysphagia	87 (39.1)	38 (57.6)	0.008

PCI, Percutanous Coronary Intervention; CABG, Coronary Artery Bypass Graft.

to repair with 32.7% of LAA patients and 44.6% of RAA patients ($P = 0.11$).

A vascular surgeon was involved in 90.1% and 75.8% of LAA and RAA cases ($P < 0.01$), respectively whereas a cardiothoracic surgeon was involved in 49.1% and 68.2% ($P < 0.01$). Rates of intraoperative complications, death within 30 days, return to the operating room, symptom relief, and endoleaks were not significantly different between the 2 cohorts (Table II). Intraoperative complications occurred in 11.0% of LAA and 4.6% of RAA patients ($P = 0.13$); 30-day mortality was 5.9% in the LAA cohort versus 1.6% in RAA ($P = 0.17$). Reintervention rates were higher after hybrid repairs compared to open procedures but did not differ by arch laterality. 15.7% of LAA and 18.8% of RAA patients who received a hybrid repair underwent reintervention, compared to 7% of LAA and 3.4% of RAA patients with open repair ($P = 0.88$). Of the 222 patients who had symptoms at the time of repair, follow-up data regarding symptom status was available for 169 (76%). Among the cohort of LAA patients, 29/47 (61.7%) had complete symptom relief, 16 (34.0%) experienced partial relief and 4.3% reported no change. Almost identically in the subgroup of RAA ASA/KD procedures, 74/122 (60.7%) reported complete symptom relief, 42 (34.4%) had partial resolution and 4.9% said no change occurred postoperatively ($P = 1$).

DISCUSSION

ASA/KD is a rare vascular anomaly and RAA patients with ASA/KD are even more uncommon than patients with LAA and ASA/KD, accounting for 23% of our multi-institutional, international study population in an operative group. The higher frequency of LAA compared to RAA presentations is consistent with the 0.05% incidence of RAA in the general population,[21] making up 10% of patients

Table II. Intraoperative and postoperative characteristics and outcomes by ASA aortic arch laterality, $N = 288$

Variable	*n* (%), left arch $N = 222$	*n* (%), right arch $N = 66$	*P* value
Operative technique			0.10
Open	72 (32.7)	29 (44.6)	0.11
Hybrid	134 (60.9)	35 (53.9)	0.35
Carotid-subclavian bypass + TEVAR	26 (11.8)	11 (16.9)	
Carotid-subclavian bypass + TEVAR + coil embolization origin ASA	53 (24.1)	10 (15.4)	
Subclavian-carotid transposition + TEVAR	19 (8.6)	5 (7.7)	
Other	36 (16.4)	7 (10.8)	
Endovascular	14 (6.4)	1 (1.5)	
Hybrid operation timing			0.39
Same operation	58 (43.3)	18 (51.4)	
Staged operation	76 (56.7)	17 (48.6)	
Vascular surgeon involved	200 (90.1)	50 (75.8)	0.003
Cardiothoracic surgeon involved	109 (49.1)	45 (68.2)	0.006
Intraoperative complication	24 (11.0)	3 (4.6)	0.13
Death within 30 days	13 (5.9)	1 (1.6)	0.17
Return to OR within 30 days	21 (9.6)	7 (11.3)	0.69
Symptom relief within 1 year			0.98
Complete	74 (60.7)	29 (61.7)	
Partial	42 (34.4)	16 (34.0)	
None	6 (4.9)	2 (4.3)	
Postoperative endoleak	30 (14.1)	7 (11.3)	0.15
Reintervention	26 (12.0)	7 (11.3)	0.88
Open	5 (7.0)	1 (3.4)	
Hybrid	21 (15.7)	6 (18.8)	

with an ASA.[22] In our cohort, RAA patients with ASA had a higher incidence of KD than LAA patients. 56% of LAA patients with ASA also have KD,[22,23] whereas 84% of RAA patients with ASA have a KD,[22] which is consistent with our findings.

The anatomic abnormalities involved in ASA/KD can lead to several complications, including life-threatening rupture or aortic dissection.[24] In a study of 312 patients from a single institution, ASA was associated with aortic pathology, including aortic aneurysm or dissection in 9% of patients with LAA and 16% of patients with RAA.[22] Patients with ASA/KD may also present with symptoms of dysphagia, dyspnea and dysphonia resulting from compression of the esophagus and trachea.[25,26] Either symptomatology or associated pathology may be an indication for operative intervention. Surgical management of ASA/KD can involve several different approaches, including both open, all endovascular and hybrid approaches, which integrates subclavian artery transposition or bypass with TEVAR.[13]

RAA patients were significantly more likely to have symptoms as an indication for intervention and presented more frequently with dysphagia. The relationship between dysphagia and tendency toward intervention is consistent with a retrospective study of 152 patients with KD, where 87 underwent no intervention and 65 received either open or endovascular repair. In this study, dysphagia was associated with an increased likelihood of operative intervention, although this was not compared by arch laterality.[18] In their surgical group, 35% had dysphagia, which is comparable to the 43% in this study's cohort. Given that RAA patients in this cohort were more likely to present with dysphagia, they trended to have surgery at 4 years younger age. Even so, there was no significant difference in symptom improvement or resolution when stratified by type of repair or by arch laterality. Notably, there was a higher incidence of KD in RAA patients in our cohort (66% in LAA, 86% in RAA, $P < 0.01$), and though not significant, average KD diameter was larger in RAA patients (38.7 mm in LAA, 49.0 mm in RAA, $P = 0.33$).

RAA patients tended to have more advanced presentations. While published case series on surgical outcomes for RAA and ASA/KD in the literature

are biased towards symptomatic presentations that necessitated intervention in the first place, descriptions of symptoms of dysphagia, dyspnea, chest pain and recurrent aspiration pneumonia are similar to the symptoms seen in our study.[17,27] The higher incidence of KD in RAA and larger size, leading to more prominent compression of the trachea and esophagus, likely explains more heterogenous symptoms in patient presentation. In the aforementioned study of 152 KD patients,[18] the authors also speculate that a larger KD size paired with the RAA may lead to increased wall shear stress, given the ''nature of the acute curvature'' of the RAA compared to the LAA, which may increase the risk of rupture or dissection and decrease the threshold for surgical intervention.

This data did not indicate any additional patient factors that may contribute to the more symptomatic presentation of ASA/KD in patients with RAA. Hypertension and concomitant arterial disease were common in both RAA and LAA groups and more prevalent than in the general population. Other authors have found hypertension to be the only comorbidity to be significantly more prevalent in KD patients who underwent surgery as opposed to the nonsurgical group, 71% vs. 55%.[18] The prevalence of hypertension is similar to the 69.8% in our reported cohort. In a study of 104 patients with KD,[28] only 6 patients had aortic pathology (including dissection, intramural hematoma, chronic aneurysmal disease and saccular aneurysm); all 6 patients had LAA and risk factors for atherosclerotic cardiovascular disease (ASCVD). The authors suggest that ASA with KD is a congenital anomaly, and the presence of hypertension and atherosclerotic disease likely leads to eventual symptomatic presentation. Interestingly, the study notes the incidence of ASCVD was significantly higher in ASA with LAA than with RAA, 56% vs. 7.4% ($P < 0.0001$), which does differ from our findings, where concomitant arterial disease was not significantly different by arch laterality. However, this is not necessarily a direct comparison as this study looked specifically at ASCVD whereas our definition of concomitant arterial disease included aortic, coronary, and peripheral artery disease, both atherosclerotic and aneurysmal. Although not statistically significant, in a study of 312 patients from a single institution, 26% of LAA versus 5% of RAA patients had aortic pathology, including aneurysm and dissection ($P = 0.21$).[22]

Hybrid technique was the favored surgical approach across the participating institutions in this study, with carotid-subclavian bypass with TEVAR and coil embolization most common in LAA patients, and carotid-subclavian bypass with TEVAR without coil embolization in RAA patients. The preference towards hybrid repair is consistent with current literature, where over recent years repairs have shifted from open repair towards hybrid and endovascular approaches, particularly in the past decade.[6,29] In a study across 7 institutions of ASA, from 2006 to 2013, 71% of 21 patients underwent hybrid repair, consisting of subclavian to carotid transposition or bypass plus TEVAR. A study of 22 patients with ASA/KD over a 15-year period showed that the frequency of endovascular repair increased after the commercialization of TEVAR in 2005 from 33% to 63%, and from 2010 to 2014, was adopted in 80% of cases.[13] In this study, there was no significant difference in surgical technique by arch laterality and choice of intervention type was more likely guided by the patient's individual anatomy, comorbidities and contraindications, and the surgeon's comfort with a particular approach.[30] However, a cardiothoracic surgeon was more likely involved in RAA cases in our study, perhaps due to the rarity and increased complexity of RAA with ASA/KD requiring a multidisciplinary approach.

The incidence of 30-day mortality in the present cohort is comparable to that observed in a meta-analysis[6] including 27 studies reporting on a total of 332 patients. They reported a pooled rate of death within 30 days after repair of 1.62% (95% confidence interval (CI) 0.05%−4.53%) in patients undergoing open or hybrid surgery through sternotomy/thoracotomy and 1.96% (95% CI 0% to 6.34%) in patients undergoing endovascular or hybrid treatment without sternotomy/thoracotomy. While this study's reported incidence of 30-day mortality is higher than the systematic review (5.9% in LAA and 1.6% in RAA), it is important to note their CI is wide and in fact, for those who did not have sternotomy/thoracotomy, encompasses this reported value of 5.9%. The pooled rates of symptom relief in the systematic review were available for 99 patients in the meta-analysis, and were 99.52% (95% CI 92.05%−100.00%) in those with sternotomy/thoracotomy and 95.79% (95% CI 83.96%−100.00%) in those without sternotomy/thoracotomy. While this study reports lower symptom relief rate (60.7% in LAA and 61.7% in RAA achieved complete relief), this study has the strength of uniformly defined and collected data, as opposed to the heterogeneity of a systematic review. Furthermore, it is possible that the symptom relief rate is lower than that of other studies because patients with recurrent symptoms, or lack of symptom relief, are more likely to follow-up than those whose symptoms resolve or improve.

Overall, rates of intraoperative complications, death within 30 days, return to the operating room, symptom relief, and endoleaks were acceptable in both groups and not significantly different. Other authors have also demonstrated that hybrid techniques are considered to be overall safe and effective.[31] In a 15-year study of 13 patients undergoing hybrid repair, 30-day mortality occurred in 1 patient, endoleaks occurred in 4 patients, and dysphagia was relieved in all but 1 patient.[13] In a study of 10 patients undergoing repair of KD between 2005 and 2010,[31] there was no in-hospital mortality, 2 patients developed endoleaks, and all patients remained free of symptoms. Although the rates of reintervention were higher in hybrid repair than open, these likely reflect endoleak repairs associated with the hybrid approach.

In this multi-institutional study approach, the operative procedures were performed by surgical teams at 20 different centers, with varying protocols. While this introduces heterogeneity into the surgical technique, these results also represent a more pragmatic evaluation of outcomes that includes tertiary referral centers and community-based practices. Similarly, given the multi-institutional standardized nature of the data collection and desire to minimize the burden of data collection, very granular details of the surgical technique were not able to be captured. Another potential limitation is the vascular focus of the paper, in the sense that the authors are predominantly vascular surgeons, and therefore data about repairs performed without the presence of a vascular surgeon could be excluded. The exclusion criteria of patients <18 may have excluded cases that were more severe and/or symptomatic that required repair prior to age 18. Additionally, due to the retrospective nature of data collection, there is inevitably missing data that cannot be obtained, particularly for the long-term outcome variables. Given the relatively small sample of LAA, further analysis comparing outcomes by approach and laterality were not performed as the sample sizes of each approach type in the LAA group would become very small.

CONCLUSION

Patients with RAA and ASA/KD are exceedingly rare. Compared to patients with LAA and ASA/KD, patients with a RAA and ASA were more likely to have a concomitant KD, and present with dysphagia, have symptoms as an indication for intervention, and be treated at a younger age. Properly selected open, hybrid and endovascular repair approaches are equally safe and effective in patients with ASA/KD, regardless of arch laterality. In patients with ASA/KD who underwent surgical repair, the majority achieve complete or partial symptom relief, regardless of arch laterality.

SUPPLEMENTARY DATA

Supplementary data to this article can be found online at https://doi.org/10.1016/j.avsg.2023.05.005.

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