Aberrant Left Subclavian Artery Arising from the Main Pulmonary Artery with Different Associations : A Case Series

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Abstract

Anomalous Left Subclavian Artery (ALSA) is a congenital anomaly, in which the left subclavian artery arises from the Main Pulmonary Artery (MPA). Patients may remain asymptomatic and may be later diagnosed either at autopsy or incidentally during angiographic studies for associated cardiac defects. The surgical management of such defect is not clearly defined. We hereby present a case series of three patients we have treated from July 2015 to January 2021, who were diagnosed incidentally during the routine investigation and treated accordingly.

Key words: Aberrant Left Subclavian Srtery (ALSA); Main pulmonary artery (MPA); Tetralogy of Fallot (TOF); Right aortic arch (RAA); Patent Ductus Arteriosus (PDA); Right Aberrant Subclavian Artery (RASA).

Introduction

One of the commonest embryological anomalies of the aortic arch is the Aberrant Right Subclavian Artery (ARSA) and in published literature this anomaly was reported as “arteria lusoria”. Hanauld, in 1735 first reported this anomaly.[1] The congenital Right Aortic Arch (RAA) is a rare aortic arch anomaly having an incidence of 0.1-0.5%.[2,3] The Aberrant Left Subclavian Artery (ALSA) is one of the RAA branches otherwise known as left lusoria artery.[3]

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It originates distal to the right subclavian artery (Last RAA branch) and often it travels behindthe oesophagus to supply the left upper extremity.[3] In comparison to the rarity of RAA, the ALSA is far rarer than the Aberrant Right Subclavian Artery (ARSA) that originates from Left Aortic Arch (LAA). Nevertheless, ALSA is more frequent in case of RAA, than ARSA in case of LAA.[4] The RAA having isolation of ALSA is another rare developmental anomaly where LSA originates exclusively from the pulmonary artery communicating via ligamentum or Ductus Arteriosum (DA) having no aortic communication.[5]

Prenatal and postmortem examinations revealed 0.5-2% incidence of ALSA in general population.[6] And the incidence rate can rise in patients with trisomy-21.[7] Right aortic arch, ALSA and kommerell diverticulum can be associated with intracardiac pathology like Fallot’s Tertralogy (TOF), Atrioventricular Septal Defect (AVSD) Ventricular Septal Defect (VSD) Persistent Left Superior Cena-Cava (PLSVC) Ductus Arteriosus, Double Outlet Right Ventricle (DORV) truncus arteriosus, mitral valvular atresia etc.[8-11]

Many surgical approaches, like primary ALSA translocation, kommerell diverticulum resection, division of ligamentum during childhood, has been suggested for the management from earlier time of this century.[12] Studies support better symptom relief and late complications prevention by this approach.[13] We hereby sharing our experiences relating tothe operative procedure in a group of three patients with aberrant left subclavian artery arising from the main pulmonary artery who were diagnosed incidentally.

Case Report 1

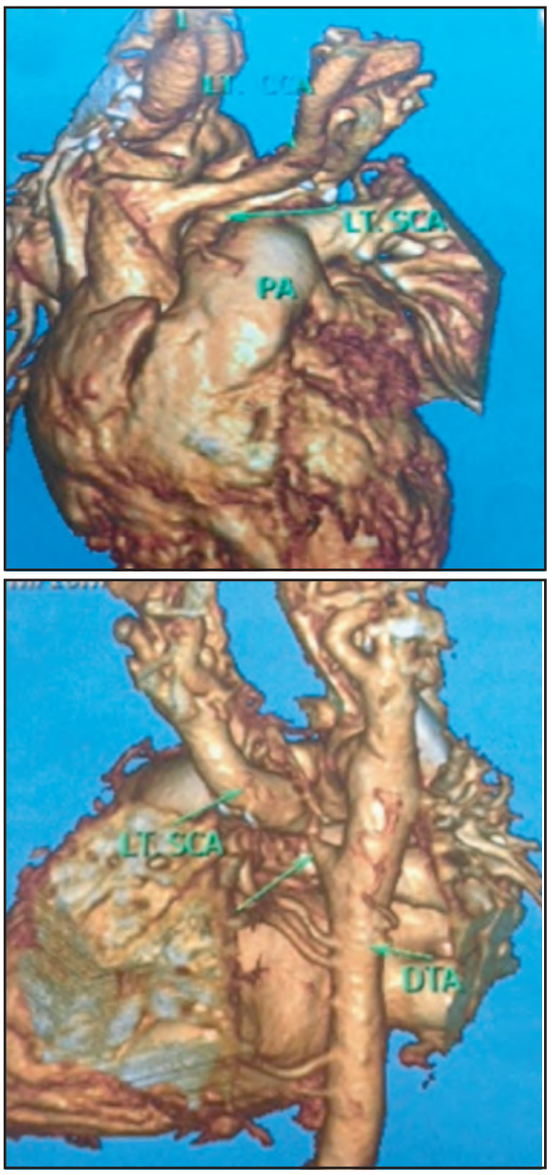

Figure 1 CT-angiogram reveals ALSCA arising from MPA

A 1½ years old male baby having congenital heart disease and product of full-term LUCS delivery was admitted at our institute with complaints of vomiting since birth after feeding. He had a history of recurrent attacks of respiratory tract infection, poor feeding, palpitation and failure to thrive. On examination, he was having mild pallor, blood pressure: left arm: 42/21 mm of Hg, right arm: 67/35 mm of Hg, Spo2: 91% (Left arm), 99% (Right arm) Pulse: feeble on left side other pulses were normal. The apex beat of the baby was placed at left 5th intercostal space just lateral to the mid-clavicular line, thrusting in nature. He was having a normal S1, but wide split was present in S2, along with a continuous murmur (3/6). Doppler echocardiography revealed dilated LA, LV and good bi-ventricular functions. Right-sided aortic arch was seen. Two vascular channels, communicating with descending thoracic aorta (Continuous collateral flow pattern in the vascular channel, but course could not be delineated) were also seen. The patient was initially taken to OR for PDA ligation through a left thoracotomy. No PDA was found per-operatively, rather we found the Left Subclavian Artery (LSA) was soft and blood was deoxygenated, but the origin was not seen from the left thoracotomy. The procedure was abandoned with a plan for definitive surgical correction after further evaluation. The chest was closed and sent the patient to ICU.

A contrast CT aortogram was done to re-evaluate the patient's heart and great vessels. It revealed situs solitus with Levocardia, right sided aortic arch. The subclavian artery was dilated and was arising from the main pulmonary artery. The left common carotid artery was arising from the ascending aorta and there was communication between the left subclavian artery and descending thoracic aorta.Multiple collateral arteries were present in the neck region and the left vertebral artery was dilated.Left ventricle was also dilated. Patient's cardiac catheterization data showed: PA pressure- 35/20/25 mm of Hg (SpO_2-80%), Left subclavian artery: 42/27/32 mm of Hg, (Spo_2-96.6), AO: 75/55/62 mm of Hg (SpO2-94 %), Qp-3.2 L, Qep- 1.6 L, Qs: 1.74 L, Qp/ Qs :1.8: 1, PVR: 5.9, SVR: 32.7, PVR/SVR: 0.18, L-R :1.6 L.

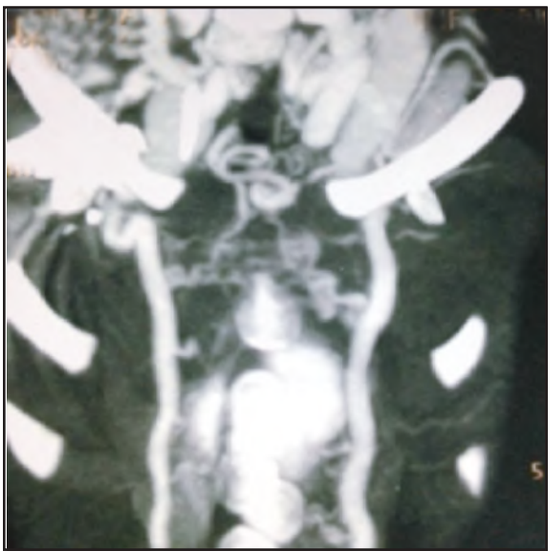

Figure 2 Multiple MAPCA in neck region

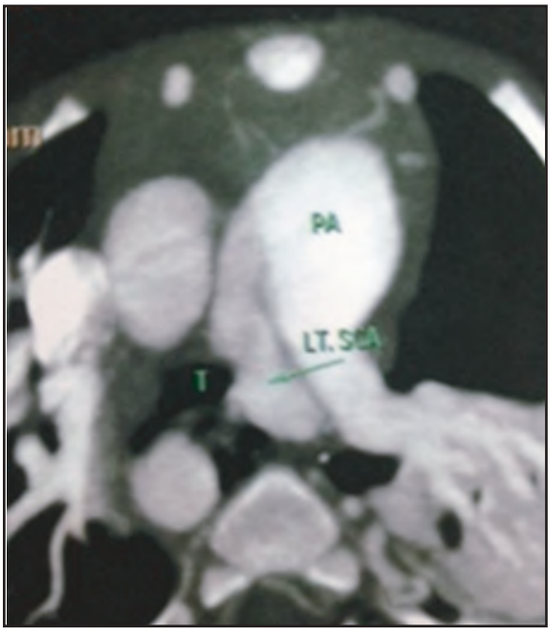

Figure 3 CT-angiogram reveals ALSCA arising from MPA

This time after proper planning patient was again taken to OR and chest was opened through median sternotomy under GA.P er-operatively thymus was absent, normally related great vessels seen. Right sided aortic arch was present, whereas left common carotid artery arises from ascending aorta, leftsubclavian artery arises from MPA in PDA position. Large conoventricular septal defect, narrow dysplastic RVOT and bicuspid pulmonary valve seen.

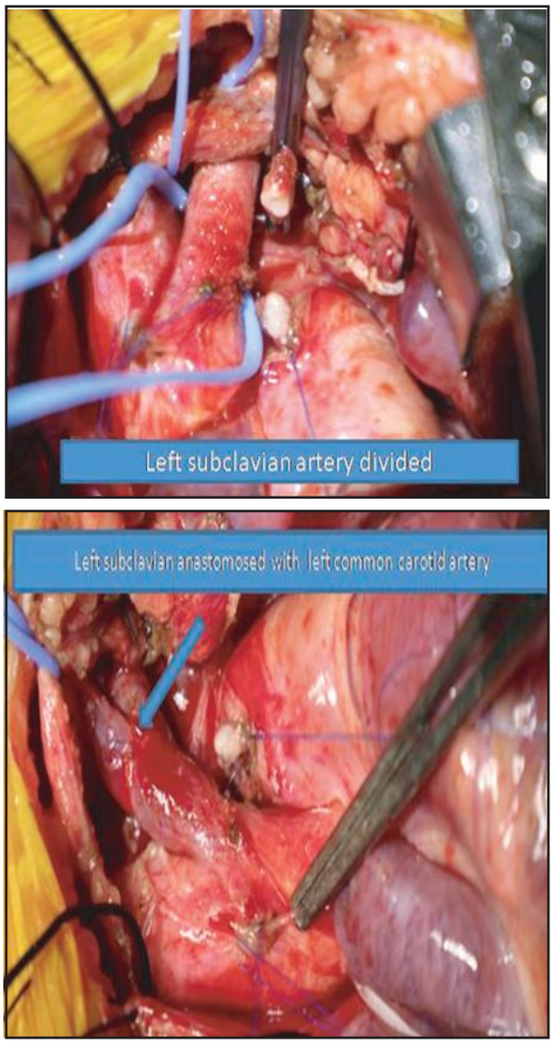

Figure 4 Anastomosis is done between LSCA and LCCA

After diagnostic confirmation, definitive surgery was planned and side to side anastomosis was done between LSA and Left Common Carotid artery (LCC). The fibrous attachment between the aorta and the aberrant subclavian artery was divided to release the trachea and oesophagus. The patient was then shifted from OR to ICU with stable hemodynamics. Postoperative CT angiogram of heart and great vessel revealed patent anastomosis between LSA and LCC. The left subclavian artery and its branches are dilated. Left ventricle was found dilated. Much reduction of collateral arteries in the neck is seen as described previously. He was discharged from the hospital on 17th POD.

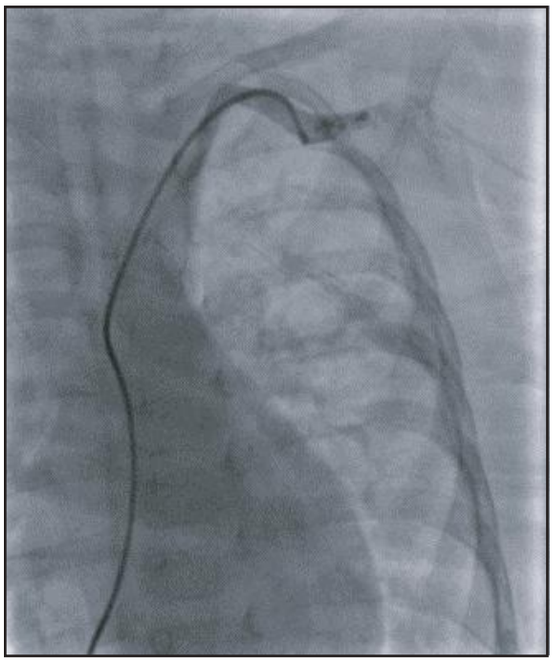

Figure 5 Cardiac catheterization showing catheter travel from RV to PA to LSA

Case Report 2

1 year 4 months old baby of consanguineous parents, one of twin preterm baby came to our hospital with the complaints of bluish coloration of skin during crying, occasional vomiting, and easy fatigability. General examination revealed, patient was cyanosed, having SpO_2 of 88% (Right hand), 92% (Left hand) and respiratory rate was 36b/min. His pulse was 96b/min (right hand), 104b/min (Left hand) and heart rate was 104 b/min, regular. Patient's blood pressure was 81/46 mm of Hg (Right hand) and 67/43 mm of Hg (Left hand).

Doppler Echocardiography revealed peri-membranous VSD with bidirectional shunt, severe valvular pulmonary stenosis, (PPG 85mm of Hg), moderate PDA, good biventricular function, right-sided aortic arch. CT angiogram showed situs solitus, levocardia, pulmonary infundibulum, and valve stenosis, hypoplastic branched LPA. Peri-membranous VSD seen. He was having right aortic arch; interrupted left subclavian artery was also seen.Patient's LCC artery was arising from the distal part of the ascending aorta. His right ventricle was hypertrophied.

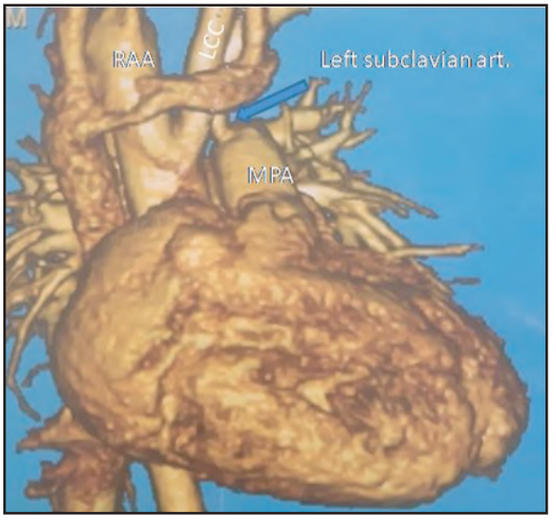

Figure 6 LCCA arising from ascending aorta

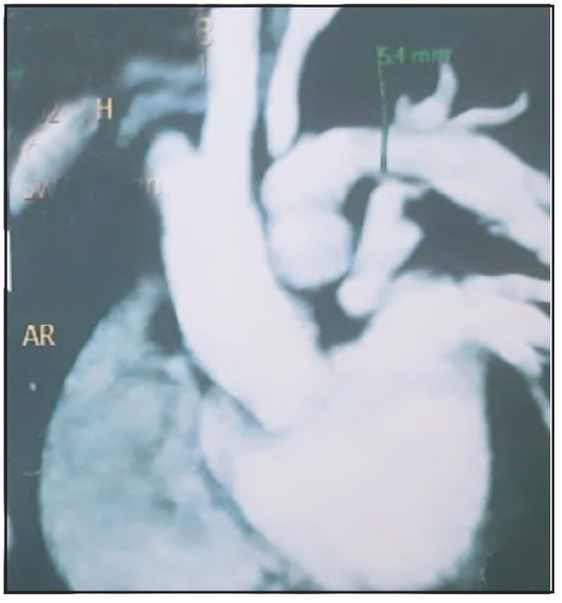

Figure 7 CT-angiogram reveals ALSA arising from MPA

During surgery we relocated the left subclavian artery and anastomosed it with LCC artery in an end to side manner. Dacron patch repair of VSD, RVOT muscle band was resection and pulmonary vulvectomy was done. We created monocusp by 0.1 mm PTFE patch and augmented the RVOT by pericardium. A fenestration was created in the IAS and the patient was weaned from CPB without any difficulties (CPB time 188 minutes, aortic cross-clamp time 128 minutes). After chest closure patient was shifted to ICU in a stable condition.

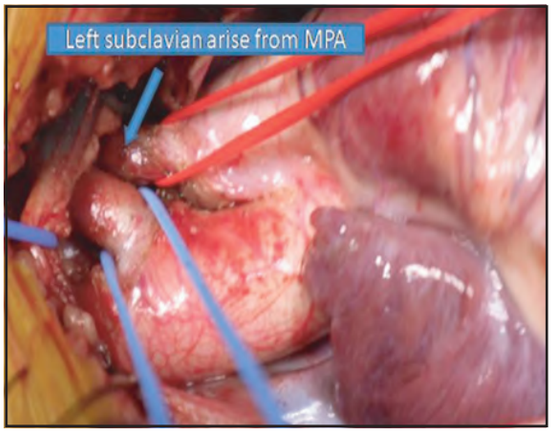

Figure 8 Peroperative finding

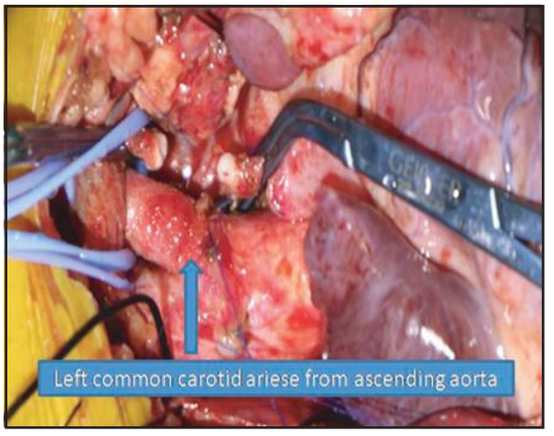

Figure 9 ALSA is transected

Case Report 3

A girl of 10 years of age was admitted in our institute with the complaints of exertional dyspnea and recurrent respiratory tract infection since birth. Examination revealed her heart rate was 96b/min and her blood pressure was 109/65 (77) mm of Hg, respiratory rate-28/min, SPo_2-93%, S1, S2 audible with systolic murmur (3/6), best heard at the left sternal border. Doppler echocardiography showed, situs solitus, levocardia, Tetralogy of Fallot, conoventricular VSD (Bidirectional shunt predominantly R-L,) severe valvular (PPG-93mmHg) and moderate infundibular PS (PPG-52mmHg), RCC prolapse with moderate AR. Mild post stenotic dilatation of MPA, LPA origin stenosis, PDA or MAPCA, left aortic arch, good biventricular functions were also seen. Cardiac catheterization revealed TOF with moderate AR. After proper planning, counselling and consent, chest was opened with median sternotomy under general anesthesia. We completed the purse-string sutures for CPB cannulation and started dissection of PDA. During dissection we identified that it was not PDA, rather it ran up and continued as the left subclavian artery. We then carried out our dissection further and we found that the left subclavian artery was missing at its normal location. Then we snagged the aberrant left subclavian artery, and checked the left radial arterial pressure. We observed that there was no pressure variation on the left hand after snagging for 3 min, so we ligated the pulmonary end of the aberrant left subclavian artery. Later on, we went to CPB and performed the required repair for TOF. She was then shifted to ICU with hemodynamic stability. Patient was weaned from mechanical ventilation on 1st POD and was discharged from hospital on 7th POD.

Study period of above 3 cases was July 2015 to January 2021. The authors have taken appropriate consent from the patient and patient attendants, that their images and other clinical information to be reported in the journal.

Discussion

The arch of the aorta with its branches starts to develop from the third gestational week. The common arterial trunk develops as a part of primitive heart and in due courseit divides into six pairs of aortic arches. The aortic arches then fuse to form bilateral dorsal aorta. Eventually after some transformations aorta proper and its branches develop. Depending on the presence or absence of the various arches, different arch anomalies can happen.[9] Right sided aortic arch occurs as a result of persistent right fourth aortic arch and regression of left aortic arch. The aortic arch in this anomaly crosses over the right main bronchus.[10] It usually has three subdivisions, a) right aortic arch, withmirror-image branching (59%), commonly associated with tetralogy of Fallot b) RAA with aberrant LSA (39%) c) RAA with an isolated LSA (1%).[14]

In case of right sided aortic arch there is increased chance of chromosomal abnormality.One study showed one third of patients with right aortic arch having chromosome 22q11.2 deletion have no cardiac anomalyand a thirdof patients have

aberrant left subclavian artery.[11] In a series of fetuses with a right aortic arch in association of TOF or common arterial trunk the incidence of chromosomal abnormality was 46%.[15] Two of our patients in this series had RAA and an ALSA that were connected with MPA at PDA position. As our patients refused chromosomal analysis the presence of chromosomal abnormality could not be ruled out,yet our 2nd case had thymic aplasia and post-operative hypocalcemia so the presence of DiGeorge syndrome (Chromosome 22q11.2 deletion) could not be confirmed. One patient had isolated aberrant left subclavian with RAA, one patient had RAA with aberrant left subclavian with TOF, and one patient had LAA with aberrant left-subclavian with TOF.

Usually, the patients with isolated LSA those produce clinical symptoms, remains dependent on the patency of the ductus. Although it was absent in our series. The isolated LSA patients frequently remains asymptomatic and, in most cases, they are identified when associated cardiac defects are present, or when a lower blood pressure is observed in the left upper limb. These patients may manifest both congenital subclavian artery steal and pulmonary artery steal syndrome. They may have vertebrobasilar insufficiency if blood flows from the left vertebrobasilar artery to the pulmonary artery (Pulmonary steal) or to the subclavian artery (subclavian steal). In case of increased left arm circulation due to left upper limb exercise, symptoms like visual disturbances, faintness, syncope or headache are usually aggravated. Patients with pulmonary steal syndrome may develop excessive pulmonary circulation.Ischemic symptoms like pain, weakness, coldness and a low length may sometime be present in the left arm.[16,17] Luetmer and Miller in a study showed patients with left arm ischemic symptoms as well as vertebrobasilar insufficiency.[16] The age of the symptomatic patients ranged from 22-53 years, and symptom duration ranged from >1-11 years before diagnosis. In our series, the age of the patient ranged from 1 year to 10 years and they have no symptom of vertebrobasilar insufficiency.

There are still lot of controversy regarding the optimal therapeutic approach. The therapy mostly encompasses simple LSA ligation, surgical re-implantation, device closure of PDA, and routine regular follow-up of the patient's condition.

Although many authors suggested simple ligation of LSA or PDA as effective therapy, but it has been shown that persistent isolated LSA if not re-implanted surgically into the aorta, provides an anatomic foundation for subclavian steal syndrome.[7] We relocated the aberrant left subclavian and anastomosed with a left common carotid artery in 2 of our cases with success,at <2 years of age. We ligated the aberrant left subclavian in the other case with success. Our recommendation is to perform corrective surgery before the vertebrobasilar insufficiency or ischemia of the arm becomes prominent, even though affected individual may clinically be asymptomatic.

Limitation

This is a case series with small sample size. A larger sample multicenter study would have brought a more representative result and would have given the readers a better understanding of the outcome.

Conclusion

There are multiple factors to be considered and a safe intervention strategy to be sorted out when aberrant subclavian artery surgery is planned. As, a single guideline which fits all the patients with this type of anomaly is yet to be present, and randomized data orconsensus on this entity are less, every diagnosed patient should be managed on its own merit. Larger multicenter study is required for standardization of the treatment.

Recommendation

These patients usually require a varied management guideline. So, we recommend early diagnosis and prompt corrective surgery for best result for the patients.

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Contribution of authors

PKB-Conception, citing references & final approval.
SD-Drafting, citing references & final approval.
NIHC- Design, citing references & final approval.
MISAM-Drafting, citing references & final approval.
SI-Conception, critical revision & final approval.
MAKS-Design, critical revision & final approval.
MS-Design, Design, critical revision & final approval.

Disclosure

All the authors declared no conflict of interest.

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