Paradigm Shift in the Management of Isolated Interrupted Aortic Arch in Adulthood

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Abstract: An interrupted aortic arch (IAA) is a rare type of congenital heart disease, and few patients survived into the adulthood. Surgical reconstruction is still the recommended approach, despite its complexity and considerable complication. In addition, patients with IAA usually suffer from several other important congenital heart anomalies, which increase the complexity of surgical management of IAA. Although endovascular therapy has replaced surgery in the treatment of the majority of non-IAA, its applicability in IAA is still matter of debate. In the present review, we have discussed about various therapeutic solutions of IAA, and present a stepwise approach for its endovascular management. (Curr Probl Cardiol 2021;46:100717.)

Introduction

A rare type of congenital heart disease is an interrupted aortic arch (IAA), which affects approximately 1.5% of patients with congenital cardiac anomalies.[1,2] An IAA is an anomaly that can be considered the most severe form of coarctation of the aorta (CoA). In an

Conflict of interest: The authors report no conflicts of interest.
Curr Probl Cardiol 2021;46:100717
0146-2806/$ – see front matter
https://doi.org/10.1016/j.cpcardiol.2020.100717

IAA, there is an anatomical and luminal disruption between the ascending and descending aorta. An IAA is a duct-dependent lesion in neonates since this is the only way for the blood flow to pass distally to the disruption.[2] Over 98% of patients with IAAs have other congenital cardiac anomalies such as patent ductus arteriosi, truncus arteriosi, single ventricles, double-outlet right ventricles, ventricular septal defects, left ventricular outflow tract obstructions (there is posterior malalignment of the conal septum in addition to the IAA, producing a ventricular septal defect as an associated lesion [80%-90%]; this malalignment might result in left ventricular outflow tract obstructions), bicuspid aortic valves, transposition of the great arteries, aberrant innominate arteries, and aortopulmonary windows.[2] Bicuspid aortic valves occur in one-third to one-half of patients.[1] Almost all cases are diagnosed in the neonatal period and present with severe congestive heart failure or cardiovascular collapse with rapid clinical deterioration.[3] If untreated, 90% of the affected infants may die in the first year of life, with the majority in the first few days.[4] In the few cases reported in adults, the presentation varies from being asymptomatic to differential blood pressure recordings in the extremities, refractory systemic arterial hypertension, headache, claudication, and congestive heart failure with its attendant complications.[3] Survival into adulthood is dependent upon the development of substantial collateral circulation. These collateral vessels are subject to atrophy, atherosclerosis, and even spontaneous rupture, resulting in secondary complications.[5]

IAA Classification

According to the Celoria and Patton classification, IAAs can be grouped into 3 types, depending on the site of the disruption[6]:

- Type A: The disruption is located distally to the left subclavian artery. This is the second most common disruption in neonates and represents approximately 13% of the cases. Type A is more frequently reported in adults (79%).
- Type B: The disruption is located between the left carotid artery and the left subclavian artery. This is the most common anomaly in neonates, representing approximately 84% of the cases. In up to 50% of the cases of type B interruption, the right subclavian artery arises aberrantly from the descending aorta below the ductus. Type B is the most frequent in neonates (53%) and is associated with DiGeorge syndrome.

- Type C: The disruption is located between the innominate artery and the left carotid artery. This is a rare type that represents approximately 3% of all cases (Fig 1).

Diagnosis

In the physical examination of patients with isolated IAAs, according to the type of interruption, differences in pulses and pressures between the left and right upper limbs or the upper and lower limbs or even absent pulses are notable.[3] Systemic hypertension is usually present, and sometimes systolic ejection murmurs are audible in the interscapular areas. Electrocardiography can show evidence of left ventricular hypertrophy.[7] Transthoracic echocardiography is the first modality for the evaluation of the function of the left and right ventricles and the aortic valve (eg, bicuspid aortic valves, aortic stenosis, and aortic insufficiency), as well as for the detection of not only left ventricular outflow tract obstructions, ventricular septal defects, patent ductus arteriosi, and any other concomitant anomalies but also the stenotic segment of the aorta (IAA type), gradients, residual widths and lengths of the interrupted segment, and collateral vessels.[8] The absence of a significant gradient in a trans-stenotic segment with abundant collateral vessels, the narrowing of the abdominal aorta, a slow systolic upstroke, and an antegrade diastolic flow on spectral Doppler suggest severe CoA or IAAs.[8] Cardiac magnetic resonance imaging and computed tomography (CT) angiography are the gold-standard tools for detecting the types and sites of IAAs, the length of the

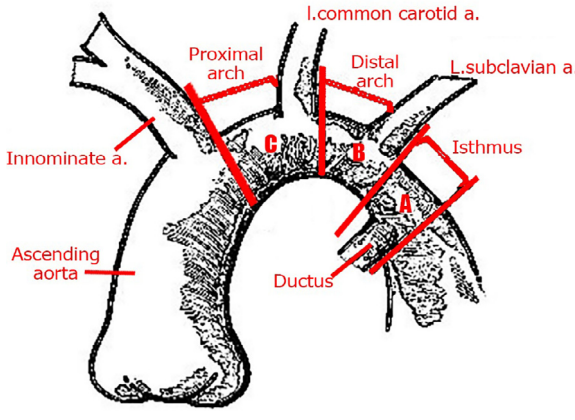

FIG 1. Interrupted aortic arch classification.

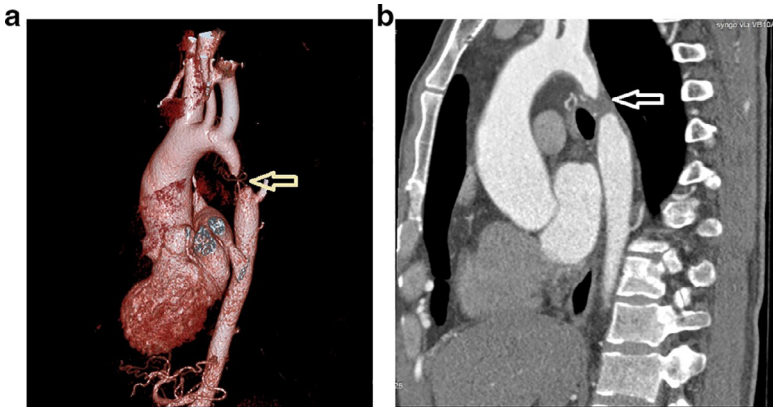

FIG 2. Interrupted aortic arch in aortic computed tomography angiography. (a) 3D reconstruction and (b) interruption site (arrow) and collaterals vessels (*).

interruption, collateral vessels, and the size of the transverse arch of the aorta at the level of the diaphragm (Fig 2 a and b).[9] They, accordingly, define the best angiographic view for the alignment of the proximal and distal ends of the interrupted segment (the most challenging part in recanalization) (Fig 3) and any vascular (origin of the subclavian arteries and their distance to the interrupted segment) and cardiac anomalies.

Treatment

The treatment of this entity in adult patients is extremely difficult, and it has not been completely established owing to a paucity of data.[10] The length of the interruption may vary from a few millimeters to larger than 1 cm. Both aortic segments, proximal and distal to the interruption, may be well-aligned or completely distorted and malaligned, which adds additional complexity to recanalization or repair. The delineation of the anatomy via imaging techniques is mandatory before treatment decisions insofar as these techniques provide an adequate planning approach.[10]

Surgical Repair

Traditionally, surgical repair has been considered the gold-standard treatment for adults with IAAs.[3] Recent advances in surgical techniques have conferred a lower risk of morbidity and mortality after surgery with more favorable long-term outcomes.[11] In adults, the longer length of the interrupted segment and the reduced elasticity of the aorta may render an end-to-end anastomosis difficult, necessitating either an interposition

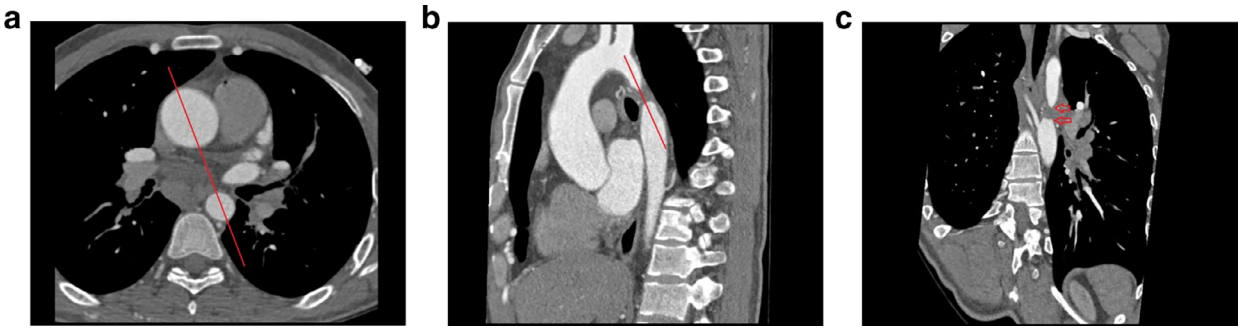

FIG 3. Preparing the working view of an interrupted aortic arch (arrow) via aortic computed tomography angiography: First, the plane is aligned on the ascending and descending aorta in the standard axial section (a), which produces a sagittal oblique projection (b). Then, the plane is aligned along the interrupted segment to produce the appropriate preprocedural projection (c).

graft or an extra-anatomical bypass.[12] An end-to-end interposition graft technique may be complicated by the need to control the collateral blood vessels; the possibility of parenchymal lung injury; damage to the recurrent laryngeal nerve, the phrenic nerves, or the esophagus; the occurrence of chylothorax; and spinal cord ischemia.[12] The most feared complication of aortic surgery is the risk of spinal cord injury and paraplegia. The risk of complications increases with prolonged aortic cross-clamp time and older age at surgery. Paraplegia is rarely encountered when using the extra-anatomic bypass technique. A side-biting aortic clamp allows the continuation of the blood flow to the intercostal arteries and in turn reduces the risk of paraplegia.[11] The use of an end-to-end interposition graft technique may increase the risk of bleeding.[12] Therefore, an extra-anatomic bypass graft instead of an end-to-end interposition graft technique appears to be safer with fewer complications and is the preferred method for surgical repair.[11] Surgical repair has some complications, the most important of which are paraplegia in approximately 0.5%-1% of cases, rebound hypertension, restenosis, and recoarctation or aneurysm formation at the site of the surgical repair.[11]

Percutaneous Reconstruction of an IAA

Transcatheter intervention is an attractive and preferred alternative to surgery for the treatment of CoA in adolescents and young adults.[13] During the long-term follow-up trials of these patients, the outcomes of transcatheter coarctoplasty were comparable to those of surgical repair, with fewer late complications.[14,15] Advances in percutaneous interventions for congenital heart disease include the percutaneous repair of CoA from straightforward luminal narrowing through to complete aortic interruption. However, the percutaneous reconstruction of an IAA is more challenging because of the tortuous course and the malaligned proximal and distal ends of the interrupted segment. A variety of methods have been drawn upon to perforate the interruption retrogradely; these methods include the Brockenbrough needle or the radiofrequency wire (Baylis).[16] With these tools, which are not steerable and usually have poor alignment with the cephalad blind end of the interruption, the incidence rates of hazardous complications such as aortic dissection, perforation, and entering the collateral vessels, which are very fragile and susceptible to perforation, have been high and failure has not been uncommon.[10] Case reports of percutaneous recanalization and stenting have been published, but there is little information on the long-term results of percutaneous treatments.

Our center is a large tertiary cardiovascular center with more than 20 years of experience in the endovascular treatment of CoA.[10,13,17] We have reported a success rate of over 98% with a mean complication rate of less than 10%.[13,17] Previously, our most common reason for the surgical referral of patients with CoA was the interrupted aorta, whereas recently, on the strength of further progress in imaging modalities (CT angiography or cardiac magnetic resonance imaging) as well as interventional equipment and techniques, transcatheter IAA reconstruction has been our preferred approach for these patients.[10] Successful endovascular experience has also significantly impacted patient prognosis. As was stated before, patients with IAAs usually suffer from various additional congenital defects needing surgical correction. A complete surgical repair (including IAA correction) is a very complex procedure that usually needs different surgical approaches and, consequently, multiple stages, which impact patient survival. In these patients, the endovascular stenting of IAAs facilitates their surgical repair and may help to simplify the surgical approach for a later repair of the remaining associated lesions.[10]

Step-by-Step Approach

Here we present our step-by-step institutional approach toward the endovascular treatment of IAAs. Of note, the percutaneous reconstruction of IAAs is usually agreed upon by the heart team's decision-making members, comprised of an adult congenital cardiologist, an interventional cardiologist, a radiologist, and an echocardiographer.

1. Full assessment of transthoracic or esophageal echocardiographic data and CT angiography including the types of IAAs; residual lumens if any; any blind ending inside the transverse and distal arch; the length of the interruption; collateral vessels; the size of the aortic arch; the best angiographic projection at the level of the diaphragm for the alignment of the proximal and distal ends of the interrupted segment, which is the most challenging part in recanalization; any vascular origin of the subclavian arteries; the distance between the left subclavian artery and the interrupted segment; and any additional cardiac anomalies. The radiological working view should also be delineated.
2. The on-call surgical team should be aware of the procedure due to such possible complications as aortic rupture.
3. Right radial artery and right femoral artery accesses are obtained for simultaneous antegrade and retrograde aortography. Simultaneous

injection is the mandatory step at the start of the procedure (Fig 4a and b). The injection should be performed in the lateral view (deep left anterior oblique projection) and more preferably, on the working view suggested by CT angiography, to exactly distinguish the proximal and distal ends and particularly delineate the projection that best shows the alignment of the interrupted aorta.

4. Due to absent or diminished femoral pulses, the arterial puncture should be performed under ultrasound guidance to ensure the cannulation of the common femoral artery. It means a central point of entry, given the size of the sheath that will be required.
5. If an IAA is confirmed, the interrupted distal end should not be crossed with a 0.035-inch guide-wire because it may lead to dissection. Depending on where the tapered end is, the retrograde or antegrade (Fig 4c and d) approach is chosen to cross the interrupted segment with dedicated instruments. In our center, the approach for crossing the interrupted segment is similar to that for chronic total occlusion revascularization with a high success rate.
6. The tapered end is chosen for the initial approach. If both ends are tapered, the antegrade approach is the preferred approach.

Antegrade Approach. In the presence of an acceptable landing zone, with the distance between the left subclavian artery and the interrupted segment more than the distal curve of the Judkins right coronary catheter or more than 10 mm, the left radial artery access is preferred. If the distance between the left subclavian artery and the interrupted segment is not satisfactory, the right radial artery approach is recommended. We prefer the left radial approach because the negotiation of the catheter through the right subclavian artery carries the risk of injuries or embolic events to the carotid arteries. Multipurpose catheters or the Judkins right coronary catheters (6F) are preferred to guiding catheters as they are more kink-resistant and also are compatible with balloons up to 2.5 mm. Additionally, the former group of catheters can be pushed firmly into the proximal blind end and with 0.014-inch chronic total occlusion coronary guide-wires, which not only have more torque control and steerability but also feature tapered tips and high tip-load. Furthermore, they easily enter the microchannels in the working projection suggested by previous imaging, usually right anterior oblique or right anterior oblique caudal. Attempts are made to perforate the blind ending with close hemodynamic monitoring, and the wire is advanced to the descending aorta (Fig 4 e and f). The stenotic segment is sequentially dilated with balloons up to 9-10 mm to allow the crossing of the catheter through the interrupted segment, and the catheter is advanced via

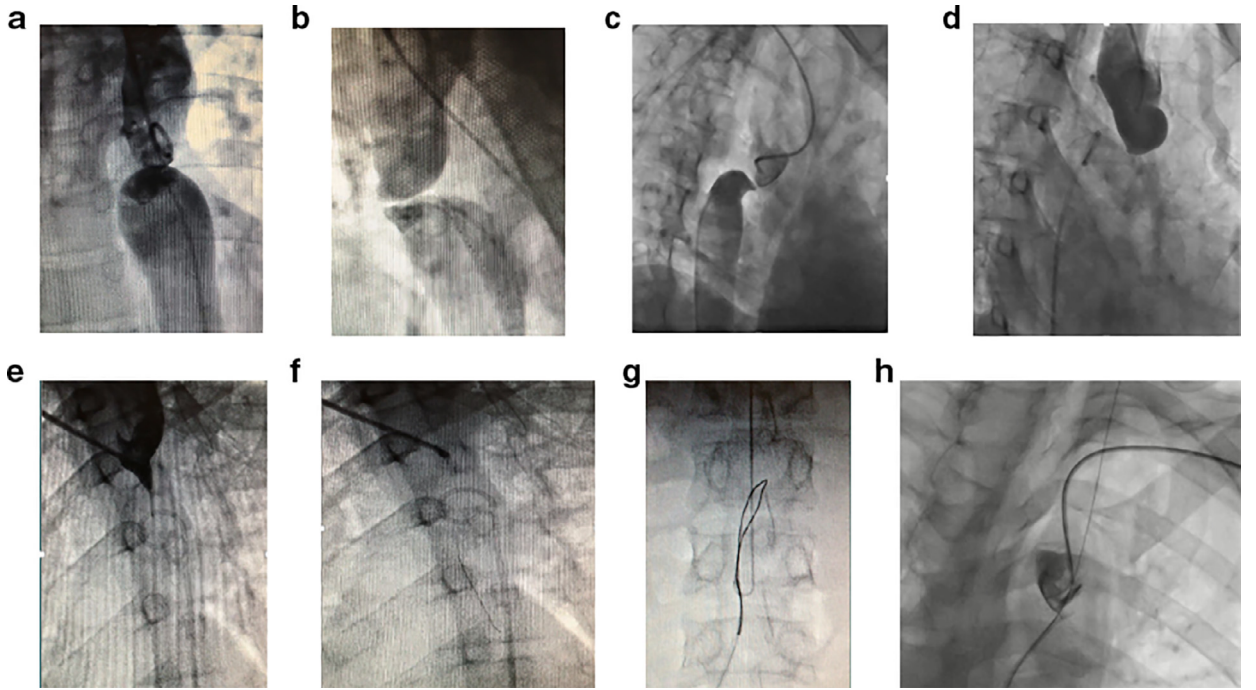

FIG 4. Step-by-step approach toward the endovascular treatment of an interrupted aortic arch: Aortography with simultaneous injection in the anteroposterior and lateral views (a and b), the identification of the proximal (c) or distal (d) tapering, catheter positioning with appropriate alignment in the tapering lesion (e), wire passage (f), wire snaring (g), and the advancement of the wire through the retrograde approach (h).

the balloon-kissing technique to the descending aorta. In all these ballooning steps, it is very important that the covered stent be available for the possibility of aortic rupture. The disappearance of the waist on the balloon confirms a compliant lesion, which is in turn amenable to stenting. If a significant waist remains on the balloon, a slightly smaller balloon is chosen and the complete expansion of the stent is postponed until a subsequent catheterization about 6 months later.

After the passage of the catheter from the coarctation site, the coronary guide-wire is substituted with a 260-cm 0.035 J tip guide-wire and snared from the femoral artery (Fig 4g). Snaring is followed by routine left heart catheterization.

Retrograde Approach. If the tapered blind ending is in the distal segment, a multipurpose catheter (6F) is pushed into the distal blind end with a chronic total occlusion coronary guide-wire in the appropriate working projections determined by previous imaging. Attempts are made to perforate the blind end, with a simultaneous antegrade aortogram in the adjacent segment to confirm the alignment of the tract. The wire is advanced to the ascending aorta (Fig 4h). Afterward, similar to the antegrade approach, the stenotic segment is sequentially dilated with increasing sized balloons to allow the crossing of the catheter through the interrupted segment. The catheter is advanced to the arch. Finally, the coronary wire is substituted with a 0.035-inch long guide-wire for the rest of the procedure.

Stenting Procedure

A long, stiff guide-wire with a soft tip (eg, a 260-cm 0.035-inch Amplatzer Super-Stiff Wire) is positioned in the ascending aorta or the right or left subclavian artery depending on the straightest wire course and the angulation of the lesion. The length of the selected stent should cover the whole length of the lesion, usually from the origin of the left subclavian artery to about 15 mm beyond the site of the CoA. The diameter of the stent (or the balloon in balloon-expandable stents) is chosen to be equal to that of the proximal isthmus at the level of the take-off of the left subclavian artery or of the distal transverse arch, whichever is larger, not exceeding the diameter of the descending aorta at the level of the diaphragm (Fig 5).

Stent Types

Both balloon-expandable and self-expandable stents have been used for the endovascular treatment of CoA. No randomized controlled trials

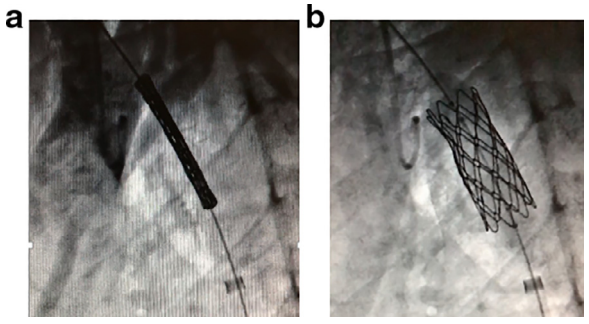

FIG 5. Interrupted aortic arch stenting: An 8-zig Cheatham-Platinum bare-metal stent mounted on an 18 × 45 mm balloon-in-balloon catheter (NuMED, USA).

have yet compared these 2 types of stents. In their cohort of patients treated endovascularly, Firouzi et al[17] showed that both stent types were safe and effective in the treatment of CoA. Notably, after propensity-score matching, Firouzi et al reported that self-expandable stents had a slightly better outcome.

A comparison between covered and non-covered stents in the non-interrupted aorta was made in a randomized controlled trial by Sohrabi et al,[18] who detected no significant superiority for covered stents. To our knowledge, no studies have compared the value of covered versus non-covered stents in patients with IAA. Due to the potential risk of dissection and microperforation during the wiring and initial ballooning, most experts have recommended covered stents in the treatment of IAAs. Covered stents have been used by our group in these patients with the following conditions:

1) Critical or subatretic obstructions, defined by a minimum diameter at the CoA site of 2-3 mm on angiography.
2) CoA associated with the atresia of the aortic lumen (interrupted).

We prefer a second (staged) postdilatation in patients with critical obstruction because of safety. This strategy avoids an abrupt and excessive increase in the diameter of the CoA site, which may result in aortic wall dissection, rupture, or aneurysm formation. The completion of dilation is undertaken after 6-12 months.

Conclusion

After improvements in transcatheter coarctoplasty and its long-term outcomes in comparison with surgical repair in adults, it is the time to

show that in adult patients with IAAs, percutaneous reconstruction, in comparison with surgical repair, may also confer great results in the hands of expert interventionists (with complete imaging evaluation and preprocedural planning, as was mentioned above) with fewer catastrophic complications such as paraplegia. In our center, the first option for the treatment of these patients is a percutaneous reconstruction, if it is anatomically suitable. The stent of choice in this group of patients is covered stents to decrease complication rates. A long-term follow-up of these patients via CT angiography or cardiac magnetic resonance imaging is mandatory to detect rarely reported restenosis, aneurysm or pseudoaneurysm formation, and stent fracture. Moreover, serial blood pressure monitoring should be considered. More studies on adult patients with IAAs are warranted to evaluate their long-term outcomes.

REFERENCES

1. Hoffman JI, Kaplan S. The incidence of congenital heart disease. *J Am Coll Cardiol* 2002;39:1890–900.
2. Varghese R, Saheed SB, Omoregbee B, Ninan B, Pavithran S, Kothandam S. Surgical repair of interrupted aortic arch and interrupted pulmonary artery. *Ann Thorac Surg* 2015;100:e139–40.
3. Ramirez Alcantara J, Mendez MD. Interrupted Aortic Arch. StatPearls. Treasure Island (FL). StatPearls Publishing Copyright © 2020; 2020 StatPearls Publishing LLC.
4. Alsoufi B, Schlosser B, McCracken C, et al. Selective management strategy of interrupted aortic arch mitigates left ventricular outflow tract obstruction risk. *J Thorac Cardiovasc Surg* 2016;151:412–20.
5. Sai Krishna C, Bhan A, Sharma S, Kiran U, Venugopal P. Interruption of aortic arch in adults: surgical experience with extra-anatomic bypass. *Tex Heart Inst J* 2005;32:147–50.
6. Celoria GC, Patton RB. Congenital absence of the aortic arch. *Am Heart J* 1959;58:407–13.
7. Hanneman K, Newman B, Chan F. Congenital variants and anomalies of the aortic arch. *Radiographics* 2017;37:32–51.
8. Goudar SP, Shah SS, Shirali GS. Echocardiography of coarctation of the aorta, aortic arch hypoplasia, and arch interruption: strategies for evaluation of the aortic arch. *Cardiol Young* 2016;26:1553–62.
9. Landeras LA, Chung JH. Congenital thoracic aortic disease. *Radiol Clin North Am* 2019;57:113–25.
10. Firouzi A, Mohebbi B, Shafiei A. Endovascular repair of interrupted aortic arch: approach with hope for fewer complications. *Arch Iranian Med* 2017;20:756–9.
11. LaPar DJ, Baird CW. Surgical considerations in interrupted aortic arch. *Semin Cardiothorac Vasc Anesth* 2018;22:278–84.
12. Grinda JM, Macé L, Dervanian P, Folliguet TA, Neveux JY. Bypass graft for complex forms of isthmic aortic coarctation in adults. *Ann Thorac Surg* 1995;60:1299–302.

13. Bassiri HA, Abdi S, Shafe O, Sarpooshi J. Early and midterm results following interventional coarctoplasty: evaluation of variables that can affect the results. *Korean Circ J* 2017;47:97–106.
14. Yang L, Chua X, Rajgor DD, Tai BC, Quek SC. A systematic review and meta-analysis of outcomes of transcatheter stent implantation for the primary treatment of native coarctation. *Int J Cardiol* 2016;223:1025–34.
15. Salcher M, Naci H, Law TJ, Kuehne T, Schubert S, Kelm M. Cardioproof Consortium. Balloon dilatation and stenting for aortic coarctation: a systematic review and meta-analysis. *Circ Cardiovasc Interv* 2016;9:e003153.
16. Tefera E, Leye M, Chanie Y, Raboisson MJ, Miró J. Percutaneous recanalization of totally occluded coarctation of the aorta in children using Brockenbrough needle and covered stents. *Ann Pediatr Cardiol* 2016;9:153–7.
17. Firoozi A, Mohebbi B, Noohi F, et al. Self-expanding versus balloon-expandable stents in patients with isthmic coarctation of the aorta. *Am J Cardiol* 2018;122:1062–7.
18. Sohrabi B, Jamshidi P, Yaghoubi A, et al. Comparison between covered and bare Cheatham-Platinum stents for endovascular treatment of patients with native postductal aortic coarctation: immediate and intermediate-term results. *JACC Cardiovasc Interv* 2014;7:416–23.