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A Systematic Review of Total Thoracic Endovascular Aortic Repair in Treatment of Kommerell Diverticulum Combined with Right-Sided Aortic Arch

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Kommerell diverticulum (KD) combined with right-sided aortic arch (RAA) and aberrant left subclavian artery (ALSA) are rare and limited to a few case reports and small series. Thoracic endovascular aortic repair (TEVAR), which is mini-invasive, is widely utilized in complicated aortic disease. We performed a systematic review of the literature to identify all patients who underwent endovascular repair for KD in terms of technical feasibility and procedural outcomes.





Published and accepted studies only in English as well as article reference lists were searched and extracted to assess case series reporting solely TEVAR in KD patients.

There were 28 patients with KD/RAA identified from 19 studies. All of them underwent endovascular technique for KD exclusion and the median age was 69 years (range 39-83 years). Hypertension (n=17) was the most common comorbidity in this cohort, followed by diabetes mellitus (n=3), hyperlipidemia (n=3), and smoking (n=3). The presenting symptoms were dysphagia (n=8, 29%), intermittent back pain (n=4, 14%), and acute aortic dissection (n=6, 21%), while asymptomatic was found in 9 patients (n=9, 32%). A technical success rate of 100% was reported associated with various managements of ALSA, proximal embolization (n=19, 68%), in-situ revascularization (n=3, 11%), and left carotid-subclavian bypass (n=3, 11%). All patients survived without severe complications and were discharged home within less than 14 days. The mean follow-up time was 9.3 months, patency was found in all patients, thrombosis and distinct shrinkage of KD aneurysm as indicated by CT-scans were noted (n=20, 71%), and type II endoleak was found in only 4 patients (n=4, 14%).

TEVAR appears to be safe and offers favorable results, but it still needs substantial evidence to support routine use in KD. TEVAR is an alternative to open repair in selected cases, but it needs further investigation in a large cohort.

Keywords: **Aorta, Thoracic • Aortic Aneurysm, Thoracic • Aortic Arch Syndromes • Embolization, Therapeutic • Endoleak • Endovascular Procedures**

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Background

Right-sided aortic arch (RAA) is rare and is found in only 0.2-2.5% of autopsy studies; it is genetically associated with 22q11 deletion [1-3]. An extremely rare type of RAA is an aberrant left subclavian artery (ALSA) originating from Kommerell diverticulum (KD), which is a persistent remnant of the fourth primitive dorsal arch, due to the aneurysmal degeneration of the origin of the aberrant subclavian artery (ASA) [4]. Specifically, in patients with KD and RAA, the left dorsal arch diminishes between the subclavian and carotid arteries so that the right subclavian artery eventually originates from the remnant right dorsal arch.

KD was first described by Dr. Friedrich Kommerell in 1936 when he performed a barium swallow examination in a patient with stomach cancer presenting dysphagia lusoria [4]. Specifically, patients with KD are usually asymptomatic, and are discovered incidentally by non-enhanced chest CT scan. When there are symptoms, the most frequent is dysphagia due to compression of the esophagus, followed by asthma and hoarseness due to tracheal compression. Other patients have a long history of intermittent pain in the chest or epigastrium, or sometimes radiating to the back [5].

Given the rarity as well as heterogeneity of RAA combined with KD, there has been no consensus on the treatment. However, Austin et al reported a 19% rate of rupture [6]. Moreover, since the presence of KD mainly predisposes to development of aneurysm and dissection, the highest rate of either rupture or dissection was reported at 53% in a long-term follow-up study [7].

Hence, due to the high rate of rupture and poor prognosis after rupture or dissection, most cardiothoracic surgeons use a surgical approach for both symptomatic and asymptomatic KD patients instead of conservative management [8]. Nonetheless, the treatment of KD varies greatly, and open repair, hybrid repair, and thoracic endovascular aortic repair (TEVAR) were all reported in recent studies [9].

However, open surgery remains a challenging procedure, with extensive incision, considerably higher need for technical dexterity, and relatively higher mortality rate (8% to 26%) [10]. Therefore, with the dramatic advent of minimally invasive procedures, TEVAR has become an attractive option for selected patients, with minimal incision and favorable results. The optimal treatment strategies of endovascular repair applied for KD patients in the present era have not been established, mainly differing in whether the ALSA is reconstructed and the application scenarios (or indications) for it [11]. In-situ fenestration by needle and radio frequency, chimney stenting, and total occlusion of ALSA via coils or plug have emerged but still remain controversial [12,13].

The aim of this study was to evaluate the early and long-term outcomes of different TEVAR strategies in treatment for KD combined with RAA through reviewing the literature, so as to provide better evidence of the efficacy of endovascular repair in treatment and enrollment of KD patients.

Material and Methods

The aim of this review was to assess all relevant studies on KD treatment by TEVAR technique. Papers published between 1990 and May 2020 were included. Searches were performed in PubMed, EMBASE, and Web of Science using the terms “Kommerell diverticulum”, “Kommerell diverticulum”, “Kommerell diverticula”, “Kommerell diverticula”, “Right-Sided Arch” in combination with “TEVAR”, “endovascular” and “endovascular repair”. Additional papers were cross-checked from reference lists of included papers.

Studies that solely reported open surgery on thoracic aorta were excluded. Studies reporting hybrid repair or two-staged operation involving ascending aortic replacement were also excluded. Studies were excluded when reporting conservative therapy or best medical treatment (BMT) without endovascular repair. Studies in which data for endovascular repair were not distinguished from data for patients undergoing total arch replacement or hybrid repair without endovascular part were excluded. Data published repeatedly by the same group were excluded; however, papers from the same authors were not excluded when demonstrating further information on different cases, measurements, and consequences. Given the rarity of KD, one-case reports and review articles were all enrolled. Data were assessed for quality independently by 2 researchers. Data were specifically collected on endovascular technique, intraoperative details, postoperative outcomes, and prognosis. Our search strategy conformed to the guidelines in the Preferred Reporting Items for Systematic Reviews (PRISMA) statement standards [14].

Results

Identified Studies and Population Characteristics

According to PRISMA protocol, 19 articles were identified from 415 articles that involved “Kommerell diverticulum” or “Kommerell diverticulum”. Studies were excluded if they did not contain any information on endovascular therapies. Also, studies were excluded when only comparing endovascular techniques or reporting hybrid repair. In terms of endovascular therapy, articles in which the classification was not KD with coexistent RAA and ALSA were excluded. Among them, mostly were case reports reporting 1 or 2 patients and 1 case

Table 1. perioperative characteristics of patients divided by symptoms.

Variables	Total KD patients N=28	Asymptomatic patients n=9	Symptomatic patients n=19	P value
Age (year)	69.1±9.9	73.3±6.7	67.1±10.5	0.229
Male	18 (64.3%)	6 (66.7%)	12 (42.9%)	0.856
Symptoms	19 (67.9%)			
Pain	10 (35.7%)	–	10 (52.6%)	–
Dysphagia	6 (21.4%)	–	6 (31.6%)	–
Hoarseness	2 (7.1%)	–	2 (10.5%)	–
Rupture	1 (3.6%)	–	1 (5.3%)	–
Smoker	3 (10.7%)	1 (11.1%)	2 (10.5%)	0.963
Hypertension	17 (60.7%)	5 (55.6%)	12 (63.2%)	0.701
DM	3 (10.7%)	1 (11.1%)	2 (10.5%)	0.963
Hyperlipidemia	3 (10.7%)	1 (11.1%)	2 (10.5%)	0.963
CRF	2 (7.1%)	0 (0%)	2 (10.5%)	0.313
COPD	2 (7.1%)	1 (11.1%)	1 (5.3%)	0.575
Coronary heart disease	2 (7.1%)	1 (11.1%)	1 (5.3%)	0.575
Depression	1 (3.6%)	1 (11.1%)	0 (0%)	0.139
PAD	1 (3.6%)	1 (11.1%)	0 (0%)	0.139
Prior stroke	1 (3.6%)	0 (0%)	1 (5.3%)	0.483
Hypothyroidism	1 (3.6%)	0 (0%)	1 (5.3%)	0.483
Mean diameter of KD (mm)	46.8±11.5	49.1±12.1	45.7±11.1	0.996
Revascularization of ALSA	6 (21.4%)	1 (11.1%)	5 (26.3%)	0.359
Embolization of ALSA	19 (67.9%)	7 (77.8%)	12 (42.9%)	0.439

Values are presented as mean±standard deviation, median or n (%). KD – Kommerell diverticulum; DM – diabetes mellitus; CRF – chronic renal failure; COPD – chronic obstructive pulmonary disease; PAD – peripheral arterial disease; ALSA – aberrant left subclavian artery. * P<0.05 compared with short duration group.

cohort containing 10 patients. We analyzed data from 28 patients who were diagnosed with RAA combined with ALSA that originated from KD, and we also assessed data on demographics, details of each patient's clinical presentation, comorbidities, pathologies, management, and follow-up [12,13,15-30].

In this review, 10 patients were female and 18 were male. The median age at presentation was 69 years (range 39-83 years). We analyzed data on comorbidities. We found that the illness frequently coexists with other chronic diseases. According to our research, the majority of patients showed more than 1 comorbidity, with hypertension being most common (n=17). Other comorbidities included diabetes mellitus (n=3), hyperlipidemia (n=3), and smoking (n=3), which are known risk factors for arterial diseases. Chronic renal failure, chronic obstructive

pulmonary diseases, and coronary heart diseases were found in 2 patients each. Depression, peripheral arterial diseases, prior stroke, and hypothyroidism were each present in 1 patient.

In terms of symptoms, 9 patients (32%) were asymptomatic and most of the other patients were mildly symptomatic. Specifically, the combination of dysphagia, hoarseness, and cough was a common 'typical' symptom due to aneurysmal compression of adjacent structures (29%). Severe chest pain was found in 6 patients who underwent acute aortic dissection (AD) as a destructive consequence of KD (21%). Furthermore, 4 patients had intermittent back pain and mild chest pain and 1 patient had a rupture, which is the most severe emergency complication of KD. Patients were classified into 2 groups depending on their symptoms. Preoperative baseline characteristics are shown in **Table 1**.

Table 2. Characteristics of Kommerell diverticulum with right-sided aortic arch via TEVAR technique in the systematic review.

Author	Year	Case(s)	Age	Gender	Symptoms	Comorbidities
Okada et al	2000	1	76	M	Hoarseness	Coronary heart diseases
Naoum et al	2008	1	58	M	Intermittent back pain	Smoking, mental diseases
Klonaris et al	2009	1	76	M	Asymptomatic	HP, DM, hyperlipidemia and COPD
Hirofumi et al	2009	1	74	M	Hoarseness	Smoking
Motoki et al	2013	1	47	F	Rupture	Nr
Silveira et al	2013	1	75	M	interscapular back pain	HP, hyperlipidemia and smoking
Anderson et al	2013	1	69	F	Intermittent back pain	HP
Zhou et al	2013	1	65	M	Severe chest pain	Nr
Hao he et al	2014	1	69	M	Severe chest pain	HP
Monaco et al	2014	1	76	M	Asymptomatic	Coronary heart diseases, chronic renal failure, COPD
Mi et al	2014	1	68	M	Intermittent back pain	Nr
Gao et al	2015	1	65	M	Asymptomatic	HP
Kazuno et al	2016	1	74	M	Dysphagia and hoarseness	HP
Hai et al	2016	1	57	F	Dysphagia	Nr
Hara et al	2017	1	76	F	Asymptomatic	Peripheral arterial diseases
Hamady et al	2017	1	78	M	Dysphagia	Coronary heart diseases, HP, hyperlipidemia, hypothyroidism
Shingaki et al	2018	1	83	F	Asymptomatic	DM
Li et al	2019	1	39	M	Severe chest pain	Nr
Anirudh et al	2019	10	71±10	5 M & 5 F	4 Asymptomatic, 3 dysphagia, 3 severe chest pain	10 HP, 1 DM, 2 prior stroke, 1 chronic renal failure
Author	Size of KD (mm)	Morphology of aorta	Type of stent graft	Treatment to ALSA	F/U time	Undesirable prognosis
Okada et al	60	KD+RAA+ occluded LSA	Handmade stent graft	None	6 weeks	None
Naoum et al	65	KD+RAA+ALSA	TAG™, Gore	Bypass	17 months	None
Klonaris et al	23	KD+RAA+ALSA+AAA	Zenith™, COOK	None	18 months	None
Hirofumi et al	42	KD+RAA+ALSA	TAG™, Gore	Coils embolization	NR	None
Motoki et al	35	KD+RAA+ALSA	TAG™, Gore	Bypass	3 months	None
Silveira et al	40	KD+RAA+ALSA	Valiant™, Medtronic	Customized branching	12 months	None

Table 2 continued. Characteristics of Kommerell diverticulum with right-sided aortic arch via TEVAR technique in the systematic review.

Author	Size of KD (mm)	Morphology of aorta	Type of stent graft	Treatment to ALSA	F/U time	Undesirable prognosis
Anderson et al	20	KD+RAA+ALSA	TAG™, Gore	Plug embolization	28 months	None
Zhou et al	Nr	AD+KD+RAA+ALSA	Talent™, Medtronic	None	18 months	Type II endoleak
Hao he et al	50	AD+KD+RAA+ALSA	Zenith™, COOK	Coils embolization	12 months	None
Monaco et al	67	KD+RAA+ALSA	AVE™, Medtronic	Plug embolization	12 months	None
Mi et al	39	KD+RAA+ALSA	Grikin™, China	Plug embolization	6 months	None
Gao et al	40	KD+RAA+ALSA	Hercules™, Microport	Plug embolization	1 month	Differences in upper limbs blood pressure
Kazuno et al	33	KD+RAA+ALSA	TAG™, Gore	Plug embolization	4 months	None
Hai et al	53	KD+RAA+ALSA	TAG™, Gore	Coils and plug embolization	NR	None
Hara et al	48	KD+RAA+ALSA	TAG™, Gore	Coils embolization	5 years	None
Hamady et al	40	KD+RAA+ALSA	Customized stent graft	Bypass	18 months	None
Shingaki et al	51	KD+RAA+ALSA	Valiant™, Medtronic	In-situ revascularization	6 months	None
Li et al	48	AD+KD+RAA+ALSA	castor™, Medtech	In-situ revascularization	6 months	None
Anirudh et al	32 ± 18	KD+RAA+ALSA (3 dissections)	NR	Coils embolization	2 months	3 type II endoleak, 1 stroke, 1 bleeding

KD – Kommerell diverticulum; RAA – right-sided aortic arch; ALSA – aberrant left subclavian artery; F – Female; M – Male; F/U – follow-up; NR – not reported; HP – hypertension; DM – diabetes mellitus; COPD – chronic obstructive pulmonary diseases.

Pathological Details

The mean diameter of KD was 46.8±11.5 mm. The mean diameter of that in asymptomatic patients was 49.1±12.1 mm, while it was 41.2±10.1 mm in 6 patients who had acute AD or rupture. One patient was not measured due to extensive intramural hemorrhage and hematoma. Acute AD was recorded in 6 patients, all categorized as Stanford type B. Chronic AD and abdominal aortic aneurysm were each found in 1 patient.

Endovascular Strategies

Endovascular repair, which is a minimally invasive and effective treatment, was utilized in all 28 patients and selected

for reconstructing KD and RAA. Percutaneous puncture of the common femoral artery (CFA) was normally performed before TEVAR, while CFA exposure was only performed for patients with severe atherosclerosis and calcification. Moreover, various management methods for ALSA were noted. First, TEVAR only without further intervention to ALSA were reported in 3 articles. Moreover, further occlusion of proximal ALSA via left brachial artery was noted in 19 patients and coils were utilized in 14 patients, while plugs were used for 5 patients. Left carotid- subclavian bypass by woven Dacron graft was found in 3 patients and in-situ revascularization through thoracic branched or customized stent grafts were noted in 2 patients. Finally, a new in-situ fenestration technique by radiofrequency and subsequent stenting were reported in 1 article. In terms

of device selection, Gore TAG™ and Medtronic Valiant™ were widely utilized for endovascular repair. Grikin and Medtech from China and COOK from the USA were also noted to be used in some individual cases.

Perioperative Findings

General anesthesia was normally performed in most articles while local anesthesia was also noted in some case with full hemodynamic monitoring. Type I endoleak mostly occurred after exclusion and was found in 5 patients. Hoarseness was noticed in 1 patient after left carotid- subclavian bypass probably due to left vocal cord paralysis and recovered after thyroplasty. Mild differences in upper-limb blood pressure were instantly noticed in most of patients after ALSA occlusion. There was no intraoperative stroke or hemorrhage, and intensive care was reported in KD patients with acute dissection. All patients were discharged within 14 days.

Mean Follow-up Time and Prognosis

The mean follow-up time was 9.3 months, ranging from 6 weeks to 5 years, and 2 patients were lost to follow-up. In this review, we found that patency was reported in all patients verified by CT-scans at last follow-up. However, type II endoleak was found in 4 patients, while none of them underwent further embolization. Thrombosis and distinct shrinkage of KD aneurysm shown by CT-scans were noted in 20 patients. The symptoms of dysphagia and hoarseness disappeared in 7 patients and were alleviated in 1. There were no migration and fracture of stent grafts in follow-up, and posterior cerebral ischemia was only found in 1 patient. No arm claudication, upper-limb dysfunction and mortality were reported. The specific details for each article are presented in [Table 2](#).

Discussion

Right-sided aortic arch, normally known as RAA, with an incidence of 0.1%, is reported in case reports and reviews with a small cohort. Kommerell diverticulum originating from aneurysmal degeneration of ASA has a prevalence of 0.04% to 0.4% based on autopsy studies, with a relatively high tendency toward rupture and dissection [4,30]. According to previous studies, KD can be categorized into 5 distinct types. The ALSA originating from RAA, left-sided aortic arch (LAA) or aorto-ductal junction were defined as 3 types. The origin of ALSA arising at the junction of RAA and the descending aorta with mirror image branching was determined as another type, while the extremely rare one involved a diverticulum at the origin of an aberrant right vertebral artery arising distal to the left subclavian artery [31].

KD has been treated by open repair for decades. Total arch replacement of RAA and aberrant left subclavian aneurysm with various exposure ways under deep hypothermic circulatory arrest (DHCA) has been widely accepted [32]. The threshold of surgical option for KD is controversial. Similar to abdominal aortic aneurysm (AAA), the maximum diameter of aneurysm determines the time for operation. Some experts advised intervening when the diameter is greater than 3 cm, while others recommended 5 cm as the point for intervention [33]. Recent research has focused on the ratio of KD diameter and its concomitant ALSA, showing that the KD did not need resection until it was 1.5 times the size of the ALSA [34]. However, KD is an embryological remnant due to failed regression and the tissue is vulnerable. Given that long-term hypertension as well as wall shear stress, KD with its abnormal morphology and angulation ruptures easily and eventually becomes AD [35]. Clearly, for symptomatic patients, troublesome symptoms caused by the compression of mediastinal structures are an indication to intervene. However, owing to a high rate of rupture and dissection, surgical repair was recommended in all KD by some groups [7].

Moreover, surgical indication is feasible when patients experience symptoms from gradual organ compression [36]. Specifically, patients might have a long history of difficulty swallowing and regurgitation due to moderate compression of the esophagus. Chest pain, chest pressure, and retrosternal burning pain in the middle of the night were also illustrated in case reports [37]. Although most patients with KD are asymptomatic, symptoms present in different ways, even in symptomatic patients, due to the correlation of ALSA and adjacent structures. Provided that the ALSA is located behind the esophagus, approximately 80% of patients present with dysphagia to solid food. In the other 2 situations, between the esophagus and trachea (15%) or anterior to the trachea (5%), the symptoms may present as dysphagia, dyspnea, or stridor [38].

Open repair (OR) has been widely used and advocated in the treatment of KD in order to not only prevent sudden rupture or dissection, but also to ameliorate the symptoms arising from moderate compression [39]. OR ordinarily involves diverticulum resection/subclavian arterial reconstruction as well as total arch replacement, using DHCA [40]. However, there still remain some limitations of OR in KD treatments. In treatment of KD using OR, thoracotomy, sternotomy, ligation, aorta cross-clamping, and DHCA are required. The whole procedure remains challenging and it requires high technical dexterity and comprehensive perioperative experience. Therefore, OR carries substantial surgical risks and complications, such as left vocal cord paralysis and right vocal cord edema, as reported by Rosu [41]. Nonetheless, the mortality was 11-25% in different reviews, while the main factors associated with death were massive pulmonary embolism, mediastinitis, hematemesis,

and fistula between KD and the esophagus, respectively [4]. Subsequently, increasing experience with TEVAR has paved the way for less invasive hybrid approaches. However, even hybrid repair (HR) still needs thoracotomy or sternotomy and replacement of the ascending aorta, which may still lead to multiple complications due to vast exposure and strict operative conditions during the long surgery [42].

Furthermore, Erben et al reported that 92% of patients with KD were found incidentally and Yu et al indicated that in asymptomatic patients with KD, the morbidity and mortality associated with conventional open intervention outweighed the potential benefit of rupture prevention [5,43]. The mortality and morbidity rates of open repair in KD were not acceptable even though KD with RAA is rare, and treatment is risky and controversial. Compared with postoperative complications like bleeding, nerve injuries, and respiratory failures in OR or HR, the TEVAR technique had illustrated favorable outcomes in aorta reinstitution.

Okada et al first reported endovascular repair in a RAA patient combined with aneurysm. The procedure showed successful isolation of KD and occlusion of ALSA without instant complications and undesirable prognosis [25]. However, TEVAR is not suitable for all KD patients and accurate preoperative clinical and anatomical evaluation is extremely critical for enrollment [15]. The optimal timing for performing TEVAR on KD involves 3 main aspects: (1) the suitability of access vessels for insertion; (2) feasible tortuosity and angulation of the aorta arch; and (3) the availability of a proximal landing zone (PLZ) [44]. The limited landing area for a graft will always lead to type I endoleak as a consequence, along with the possibility of esophagus compression by stents [37]. Therefore, a sufficient PLZ is essential for the success of TEVAR in KD treatment. PLZ was proposed and classified to 3 types according to the modified Hishimaru aortic arch classification. Tinelli et al suggested that a standard thoracic stent graft was feasible for implantation when the PLZ comprised at least 2 cm of healthy aorta [44].

According to the results reported above, we found that there were no differences in the mean diameter of KD in asymptomatic and symptomatic patients and no significant difference in mild symptomatic and acute dissection or rupture. Current treatment guidelines for KD are based on a small case series of patients with no clear consensus on the surgical indications. One of the most consistent indications emphasize size, claiming that the KD aneurysm and aortic diameter need to be larger than 3 cm and 5 cm, respectively [33]. However, we found that the size of the KD was not the threshold for intervention because it would lead to disastrous consequences regardless of diameter.

There has been considerable controversy in the literature regarding the feasibility for TEVAR in KD patients. Ota et al argued that there is no volume reduction of the diverticulum after complete exclusion by stent graft, and compression symptoms cannot be attenuated [39]. However, thrombosis and shrinkage of a saccular aneurysm were widely found in this review, so that the compression by KD and corresponding symptoms would be largely alleviated [23]. Also, El Batti et al found that there was ample room between the KD and the esophagus during hybrid repair, which means that the most important aim for KD treatment was preventing rupture, although we assume that a very large KD compression may be a problem [45]. Moreover, Ota et al claimed that a “sharp angle” between the distal arch and descending aorta was an anatomical challenge for transition [39]. It may be taken into consideration in early stage of TEVAR, yet we believe that it is no longer a contraindication due to materials innovation. The introducer sheath of the new stent graft produced by Cordis (USA) is 14-F, with more flexibility compared to the normal 18-F and 24-F sheaths.

Furthermore, no matter which devices are used to treat ALSA, there remain 2 basic techniques: proximal exclusion with/without blood flow reconstruction. Percutaneous coil embolization of the proximal segment of the ALSA was performed by coils or plugs after deployment of the device in some cases [16,21]. However, some researchers were concerned that intentional coverage of the ALSA during TEVAR would increase the risk of upper-extremity subclavian steal syndrome, or possibly of spinal cord ischemia. Rizvi et al reported the need for revascularization need in investigations of cerebral and vertebrobasilar circulation [46]. To avoid arm claudication and vertebrobasilar ischemia, in-situ revascularization and bypass were performed for fear of subclavian steal syndrome [23,24]. However, based on limited cases of KD, more research and observations are needed before reaching a consensus.

Conclusions

KD, usually combined with right-sided aortic arch, is extremely rare and associated with ASA degeneration. Symptoms can be atypical for aneurysms due to persistent compression to adjacent tissues in the mediastinum. Several types of therapies were utilized and reported in the last 50 years, including best medication only, which was not recommended recently, and total arch replacement and debranching of the aortic arch followed with endovascular procedures and total TEVAR. Total arch replacement and debranching for KD under deep hypothermic circulatory arrest are not always required except for severe symptoms or lesions involving the ascending aorta. However, given the increased complexity of and limited experience with conventional surgery in such rare anatomic

configurations, with the availability of PLZ, TEVAR is more feasible for KD patients who are asymptomatic or mildly symptomatic. Even in patients who also have acute dissection or rupture, TEVAR still has an acceptable safety profile, and there have been reports of encouraging postoperative outcomes. As the technology continues to improve, TEVAR may become the ideal interventional approach for KD under conditions of

preoperative planning, careful delineation of anatomy, and strict patient enrollment.

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