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Review

Anomalous interarterial left coronary artery: An evidence based systematic overview

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Abstract

Background: Isolated anomalous left main coronary artery (ALMCA) from the right aortic sinus of Valsalva (RASV) with an interarterial course between the pulmonary trunk and aorta is a rare congenital abnormality. We performed an evidence based systematic overview spanning 4 decades to assess the prevalence, clinical features and management of this anomaly.
Methods: A computerized search spanning 40 years was conducted to identify articles describing cases of ALMCA arising from the RASV with an interarterial course. The bibliographies of all relevant articles were also searched.
Results: The search identified 264 cases. Age ranged from 3.5 months to 87 years. Male/female ratio was 2.9/1. Forty-nine percent of the cases were diagnosed postmortem. Cardiac catheterization was the most common diagnostic tool (41.7%) followed by echocardiography, magnetic resonance imaging (MRI) and computerized assisted tomography. Fifty-seven (21.6%) cases underwent surgical procedures with no mortality and low morbidity.
Conclusions: ALMCA from the RASV is associated with increased risk of sudden death, notably in young patients. Unfortunately the majority are diagnosed postmortem. More than a third present with sudden cardiac death. Echocardiography, computerized assisted tomography and cardiac MRI are valuable non-invasive diagnostic tools. Cardiac catheterization provides a definitive diagnosis in the majority. Surgical correction is the mainstay of treatment with low risk and good anatomic and functional results.

Keywords: Coronary vessel anomalies; Sinus of Valsalva; Cardiovascular surgical procedures

1. Introduction

Congenital anomalies of the origin and course of the coronary arteries are an uncommon but important cause of chest pain and sudden cardiac death. The incidence is between 0.6% and 1.3% of the population [1]. Anomalous coronary arteries are classified into seven categories according to the Society of Thoracic Surgeons [2].

In 1974, Cheitlin et al. described clinical and necropsy findings in 13 patients in whom the anomalous left main coronary artery (ALMCA) arose from the right aortic sinus of Valsalva and coursed between the aorta and pulmonary trunk and called attention to the frequency of sudden cardiac death as a consequence of this "not so minor congenital anomaly" [3]. This anomaly is rare, accounting for only 1.3% of all coronary anomalies. It is an important cause of sudden cardiac death in young people, especially during or immediately post-exercise [4–6].

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ALMCA arising from the right aortic sinus of Valsalva may be further classified based on their course relative to the great vessels into: a) septal course (through the ventricular septum beneath the right ventricular infundibulum), b) anterior to the aorta, c) posterior to the aorta, d) interarterial course (between the aorta and pulmonary trunk) or e) mixed [2]. The septal course is the most common and is generally asymptomatic. An anterior course is also usually benign, although chest pain and myocardial infarction can occur. The posterior course has been associated with syncope and myocardial infarction. The interarterial course is the most symptomatic and fatal variant [7]. In addition to an interarterial course, three other high-risk features of an anomalous artery have been identified: slit-like ostium, acute-angle take-off and intramural aortic segment. These features likely increase the risk of sudden death by limiting coronary blood flow yet further [3].

We set up a collaborative group to review individual patient data from articles describing the morbidity, mortality, clinical presentation and demographics of ALMCA arising from the right aortic sinus of Valsalva with an interarterial course between the pulmonary trunk and aorta. The results of this evidence based overview should help guide the safety and efficacy of different therapeutic strategies in the treatment of this fatal coronary anomaly.

2. Materials and methods

2.1. Search strategy

We performed a systematic search of the English-language literature published between January 1st 1966 and December 31st 2006 from the following databases: MEDLINE, EMBASE, Web of Science and the Cochrane Database. We used the search terms of "coronary vessel anomalies", "sinus of Valsalva", "pulmonary trunk or artery", "aorta", "anomalous, aberrant or ectopic coronary artery" and "interarterial course". The computer search was augmented by manual searches of reference lists from the individual reports, case series, review articles, meta-analyses and consensus statements. An independent search by a qualified librarian supplemented our search. Articles were identified as being relevant by *a priori* criteria (*i.e.* randomized trials, cohort studies, case series or case reports). Thirty eight articles were identified in the initial search, 118 articles were added by scanning reference lists. Of these, 75 articles were deemed relevant based on the selection criteria below.

2.2. Selection criteria

We included only studies that met the following criteria: cases of ALMCA arising from the right aortic sinus of Valsalva

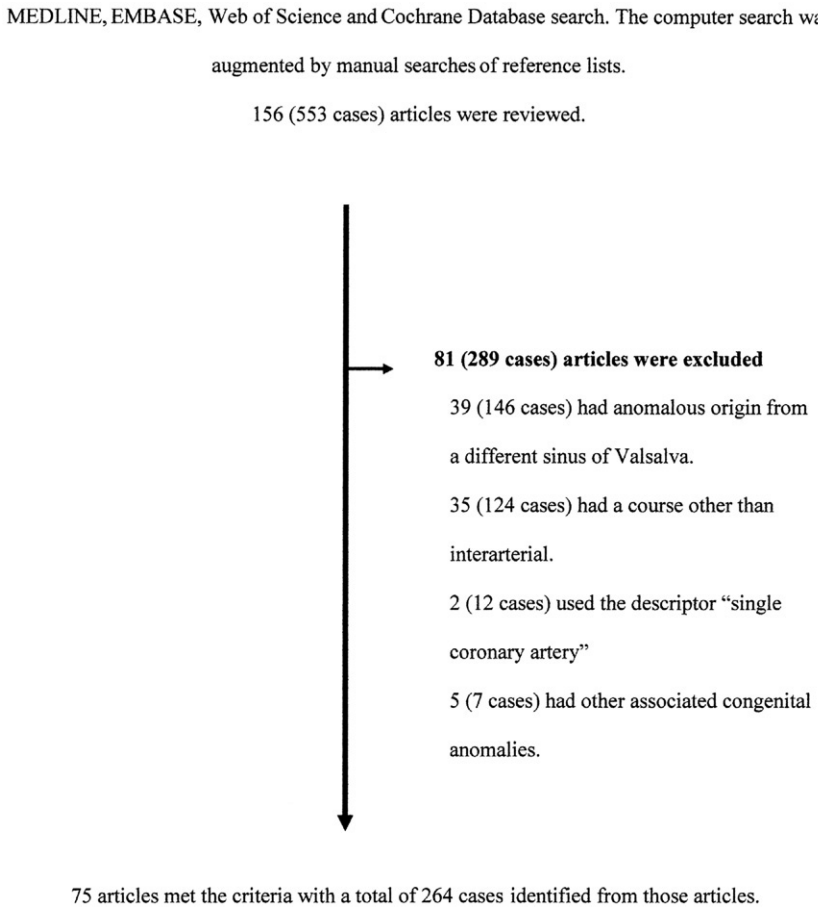

Fig. 1. Results of literature search and review.

with an interarterial course between the pulmonary trunk and aorta. Exclusions included 1) anomalous coronary artery origin from left or non-coronary sinus of Valsalva, 2) a course other than interarterial, 3) cases using the descriptor "single coronary artery" and 4) other associated congenital anomalies.

2.3. Data extraction

Relevant papers that met *a priori* criteria were identified by consensus. Outcome data were independently extracted by 3 reviewers (FM, MM, SM) and disagreements were resolved by consensus. Major outcomes were prognosis and outcome after different treatment strategies.

Frequencies or percentages were used to describe categorical variables. Continuous variables were described as mean ± standard deviation (SD) or median. All analyses were performed using JMP statistical software, version 5.2 (SAS Institute Inc., Cary, NC).

3. Results

The ALMCA prevalence and management have been assessed in non-randomized (case reports, case series and review articles) studies only. A total of 264 cases were identified [3–77] and the results are shown (Fig. 1, Tables 1–4).

Table 1 shows the demographics, clinical characteristics, diagnostic tools and ostial anatomy of the systematic review patients. Median age was 22 years and ranged from 3.5 months to 87 years (mean±SD = 35.5 ± 21.7 years). Age was not reported in 80 cases. Standard 12-lead electrocardiograms (ECGs) were performed in 53 (20.1%) and were abnormal in 21 (7.9%) patients. ECG abnormalities included ST-T wave changes (n = 9), evidence of acute or old myocardial infarction (n = 5), left ventricular hypertrophy (n = 2), ventricular tachycardia (n = 2), ventricular premature beats (n = 2) and right bundle branch block (n = 1). Standard chest X-ray was performed in 6 (2.3%) and showed pulmonary edema in 3 (1.1%) patients. A maximal stress ECG test was available for 23 (8.7%) and was positive for ischemia in 12 (4.5%) patients. Nuclear imaging tests were available for 11 (4.2%) and were positive for ischemia in 8 (3.0%) patients. Confirmatory diagnostic imaging tools are listed in Table 1.

Table 2 shows all deceased patients. Ninety-six (96/130) patients had sudden unexplained death either at rest or

Table 1
Patient demographics, clinical characteristics, diagnostic tools and ostial anatomy

	No. (%)
Gender	
Male	121 (45.8)
Female	42 (15.9)
Not reported	101 (38.3)
Presentation	
Sudden death	96 (36.4)
Chest pain	74 (28)
Syncope, presyncope	38 (14.4)
Dyspnea	12 (4.5)
Palpitations	11 (4.2)
Myocardial infarction	8 (3)
Asymptomatic	52 (19.7)
Not reported	98 (37.1)
Diagnostic tools	
Autopsy	130 (49.2)
Cardiac catheterization	110 (41.7)
Transthoracic echocardiography	24 (9.1)
Magnetic resonance imaging	13 (4.9)
Multidetector and electron beam computerized assisted tomography	9 (3.4)
Transesophageal echocardiography	8 (3)
Ostium of ALMCA	
Separate ostium	59 (22.3)
Common with right coronary artery ostium	33 (12.5)
Not reported	172 (65.2)

ALMCA: Anomalous left main coronary artery.

Table 2
Causes of death

	No. (%)
1 — Sudden death	96 (36.4)
At rest	43 (16.3)
During or after exertion	53 (20.1)
2 — Death due to cardiac causes	16 (6.1)
Coronary artery disease	12 (4.5)
Congestive heart failure	1 (0.38)
Aortic stenosis	1 (0.38)
Aortic insufficiency. Subacute bacterial endocarditis	1 (0.38)
Dilated cardiomyopathy	1 (0.38)
3 — Death due to non-cardiac causes	18 (6.8)
Cancer	4 (1.52)
Pulmonary emboli	3 (1.14)
Trauma	2 (0.76)
Liver cirrhosis	2 (0.76)
Alcoholism	1 (0.38)
Meningitis	1 (0.38)
Thromboemboli	1 (0.38)
Cerebrovascular accidents	1 (0.38)
Umbilical hernia surgery	1 (0.38)
Tuberculosis	1 (0.38)
Hemorrhage	1 (0.38)

Table 3
Treatment and outcome

	No. (%)	Number of patients completed follow-up	Range of follow-up	Complications
				No. (%)
Medical treatment	11 (4.2)	4/11	2 months–7 years (average 32 months)	0 (0)
Percutaneous coronary angioplasty	4 (1.52)	3/4	6 weeks–12 months (average 8.5 months)	0 (0)
Surgical treatment	57 (21.6)	23/57	4 months–10 years (average 18.7 months)	1/57 (1.8) severe AI

AI: aortic insufficiency.
Treatment was not reported in 62 (23.5%) cases.

Table 4
Different surgical techniques and their outcomes

Surgical procedures	No. (%)	Range of follow-up (months)	Complications No. (%)
Coronary artery bypass graft	22/57 (38.6)	4–18 (average 10) Not reported in 11 cases	0/11 (0)
Unroofing	16/57 (28.1)	4–69 (average 23) Not reported in 5 cases	1/11 (9.1) severe AI
Ostial splitting, neo-ostial creation and ostium sphincteroplasty	9/57 (15.8)	5–120 (average 22) Not reported in 2 cases	0/7 (0)
Reimplantation	2/57 (3.5)	12–24 (average 18)	0/2 (0)
Patch augmentation	2/57 (3.5)	18	0/2 (0)
Surgical angioplasty	1/57 (1.8)	Not reported	Not reported

AI: aortic insufficiency.
No surgical details in 5/57 cases.

related to exercise. At autopsy, the anomalous coronary artery with or without ischemic damage within the related myocardium, in the absence of significant coronary atherosclerosis was identified. Sixteen patients died due to known underlying cardiovascular disease unrelated to the coronary artery anomaly and the remaining 18 patients died of a pre-existing non-cardiac medical condition.

3.1. Therapeutic options and outcome

3.1.1. Surgical repair

Tables 3 and 4 show surgical techniques and their outcome. Median age was 20 years and ranged from 9 to 80 years (mean ± SD = 30.7 ± 20.0 years). Symptoms included: chest pain (n = 44), syncope/presyncope and palpitations (n = 7), shortness of breath (n = 1) and acute myocardial infarction (n = 1). Four patients were asymptomatic. The anomalous coronary origin was confirmed by surgical inspection in all cases. All patients underwent preoperative cardiac catheterization. In addition, the origin and course of the ALMCA were suspected by echocardiography in 13, computerized assisted tomography and magnetic resonance imaging in 6 patients.

Twenty-two patients underwent coronary artery bypass graft surgery to the left anterior descending or diagonal ± left circumflex coronary arteries. The internal mammary artery was the sole graft in the majority (14/22), in combination with a saphenous venous graft in 4 and the saphenous venous graft was the sole graft in the remaining 4 cases. Four patients had, in addition to the anomaly, diffuse atherosclerotic coronary artery disease (CAD) and underwent a multivessel coronary artery bypass graft. One patient underwent a combined coronary artery bypass graft with aortic valve replacement secondary to severe aortic insufficiency (AI) unrelated to the coronary anomaly. Another patient had a saphenous venous graft to the obtuse marginal branch of the left circumflex combined with arterialization of the posterior cardiac vein by anastomosing to the aorta using saphenous venous graft due to small caliber of the left anterior descending [16]. All patients were asymptomatic till the end of follow-up. Non-invasive tests (resting and stress ECG, resting and stress echocardiogram and nuclear imaging studies) showed no abnormalities.

Sixteen patients underwent an unroofing procedure. A classic unroofing procedure was performed on 12 patients in which the intramural segment was unroofed by excising the common wall between the anomalous coronary artery and the aorta until the coronary exited the aorta. In 4 patients, the affected coronary commissure in the area of the unroofing required resuspension because of disruption of the commissure by the unroofing incision (modified technique). Two patients required combined unroofing procedure with coronary artery bypass graft due to diffuse atherosclerotic CAD. Another patient required aortoplasty together with the unroofing procedure. In patients who underwent the modified technique, AI was found to be trivial in 3 and severe in 1 patient by transthoracic echocardiography at follow-up. The patient with severe AI had the Ross procedure performed 44 months postoperatively. The remaining patients were asymptomatic at follow-up with no abnormalities detected by non-invasive tests.

Five patients had an ostial splitting procedure performed. The ostium was split open and the incision was extended above the intercoronary commissure, along the intramural segment of the ALMCA to the midpoint of the left coronary sinus. Three patients had a neo-ostium created without disruption of the commissure or significant unroofing of the intramural segment of the ALMCA. One patient had the left coronary ostium enlarged by cutting the inferolateral lip and performing a sphincteroplasty on this orifice. Two patients had their ALMCA reimplanted, with a cuff of aortic wall, to the left coronary sinus combined with aortic valve replacement (n = 1) due to severe AI. Two patients underwent patch augmentation of their ALMCA ostia as the coronary artery could not be mobilized for reimplantation because of its intramyocardial course. One patient had surgical angioplasty to the ALMCA with autologous pericardium. All patients were asymptomatic till the end of follow-up and non-invasive tests showed no abnormalities.

3.1.2. Percutaneous coronary angioplasty

Percutaneous coronary angioplasty was performed in 4 patients; their ages were 43, 52, 61 and 75 years. Three patients presented with chest pain (not reported in the 4th patient). The anomalous coronary origin and course were suspected by magnetic resonance imaging in 1, transesophageal echocardiography in 2 and computerized assisted tomography in 1 and confirmed by cardiac catheterization in all patients. Two patients had intracoronary stents deployed in the ALMCA and another 2 had percutaneous coronary angioplasty to the left anterior descending and obtuse marginal branch respectively

without stent implantation. Three patients were asymptomatic till the end of follow-up and non-invasive tests showed no abnormalities (Table 3).

3.1.3. Medical treatment

Eleven patients were treated medically (3/11 refused surgery). Median age was 18 and age ranged from 4 to 53 years. Two patients were asymptomatic. The majority (6) presented with chest pain. One patient had aortic valve replacement due to severe aortic stenosis without repair of the ALMCA. Four patients were asymptomatic till the end of follow-up and non-invasive testing showed no evidence of ischemia (Table 3).

4. Discussion

The anatomic course of anomalous coronary arteries is crucial because it relates to morbidity and mortality. Autopsy series have shown that some patients who die exhibit myocardial ischemia; others have no evidence of ischemic injury and may have experienced lethal arrhythmia. The pathophysiology of compromised coronary blood flow has several putative mechanisms: (1) compression of the intramural segment of the coronary artery between the pulmonary artery and aorta; (2) a slit-like orifice to this anomalous artery with increased aortic pressure; (3) an acute-angle take-off of the coronary artery; and (4) stenosis in the intramural segment of the coronary artery, especially at the level of the commissure. These features likely increase the risk of sudden death. A slit-like ostium and acute-angle take-off narrow lumen diameter may lead to vessel collapse. Likewise, an intramural segment may be compressed during ventricular systolic contraction [3,44,46,74]. Attention to the ostium is important for optimal therapy.

Although coronary angiography is the gold standard for the evaluation of CAD, it may not completely delineate the origin or course of the anomalous coronary artery. The presence of the "dot" sign anterior to the aorta suggests an interarterial course of a coronary artery arising from the contralateral cusp [78]. In some cases, it is difficult to engage the ostium of the anomalous coronary artery because of a slit-like orifice or a membrane across the ostium. Misdiagnosis of the proximal course of the anomalous artery can occur in up to 50% of patients [5,8]. In several studies, transesophageal echocardiography has been useful in confirming the origin, proximal course and the relationship of the anomalous artery to the great vessels. It can reduce radiation time from prolonged and difficult coronary angiography and may sometimes achieve better results [41,48]. In children, the diagnosis is most frequently made with transthoracic echocardiography. With improvements in ultrasound technology, transthoracic echocardiography now allows delineation of coronary artery anatomy in adolescents and adults utilizing color Doppler flow mapping [18,22,40,49]. Novel techniques such as cardiac magnetic resonance imaging and multidetector computerized assisted tomography play an important complementary role in many patients when the results of coronary angiography are equivocal [11,12,35,36].

4.1. Surgical therapy

In symptomatic patients with ALMCA, prompt surgical repair is indicated. Management of the asymptomatic patient remains controversial although studies to assess coronary perfusion with exercise may provide additional information. The risk of sudden death or cardiac ischemia must be weighed against the risk of the operation and the risk of potential late complications such as neo-ostial stenosis, graft attrition or need for a second bypass surgery in the future. In asymptomatic patients, there is a consensus that surgical intervention is indicated to prevent the risk of sudden death but timing is controversial. Some authors recommend delaying elective surgical repair until late puberty or approximately 10 years. Others suggest elective surgical repair at less than 30 years of age. If surgical repair is declined, avoidance of strenuous physical activity and competitive athletics should be prescribed [1].

Coronary artery bypass grafting is the treatment of choice in relieving ischemic symptoms; however, long-term results may be less favorable [7]. Vein conduits to minimally diseased vessels show a patency rate similar to that of patients undergoing revascularization for atherosclerotic disease but accelerated atherosclerosis in the native vessel may ensue. Vein conduits to non-atherosclerotic vessels show poor results because of competitive flow between the vein graft and the unobstructed native artery [79]. Long-term patency of internal mammary artery grafts exceeds that of saphenous venous grafts. It also eliminates the need to open the aorta and manipulate the intercoronary commissure but subjects the patient to a bypass graft and the potential need for reintervention. Because flow through the anomalous coronary artery is minimally obstructed at rest, an internal mammary artery bypass graft may have decreased patency secondary to competitive flow. This has led some authors to recommend ligation of the coronary artery proximal to the insertion of the graft [80]. Selective arterialization of the coronary venous system may be considered in cases of diffuse atherosclerotic CAD not amenable to traditional revascularization strategies [16]. Coronary artery bypass grafting may be an acceptable therapy in an elderly patient with co-existent CAD, but commits the patient to grafted CAD and its associated risks. Patients treated with coronary artery bypass graft require long-term follow-up like any patient with CAD post-revascularization [1].

A more anatomic approach is to return the coronary artery orifice to the appropriate sinus where it exits from the aortic wall at right angles. This can be accomplished by excision and reimplantation [39] or by using a modified unroofing technique [21,63,81]. Patients require long-term follow-up to detect potential ostial stenosis and AI.

The unroofing procedure was initially described with the origin of the ALMCA at a level cephalad to the commissure. This technique has several advantages over other coronary repair techniques: 1) it relieves any ostial stenosis at the origin of the anomalous coronary by unroofing the common

wall between the aorta and anomalous coronary and relocating the coronary ostia to the appropriate sinus; 2) it creates a large neo-orifice of the anomalous coronary in the appropriate sinus that arises perpendicularly, rather than obliquely, from the aortic root; and 3) it removes the intramural segment of the anomalous coronary. This technique creates a permanent, unobstructed neo-orifice without the risk of late ostial stenosis because no circumferential suture line is placed and is ideally suited for the patient with an intramural course of the anomalous coronary. The traditional techniques of unroofing have the disadvantage of temporary detachment of the intercoronary commissure with inherent risk of post-operative AI which can be avoided by using the modified technique [21,63,81].

Reimplantation has been successful, but the coronary artery must be long enough to move directly, or a button of aortic wall must be excised with the coronary artery reimplanted in the appropriate sinus. It has the advantage of leaving the suspension of the aortic valve untouched and reestablishing the normal anatomic situation. This necessitates reconstruction of the aorta where the button was excised. Reimplantation may necessitate the reconstruction of the commissural attachments as well [1,39].

Patch augmentation can be performed as an alternative in cases of ALMCA with intramyocardial course in which neither unroofing nor reimplantation is possible because of the fixed and remote nature of the anomalous vessel [18,22]. In patients in whom the intramural course is at or below the level of the commissural attachments of the aortic valve, a new ostium of the coronary artery could be created in the appropriate sinus without disturbing the intercoronary commissure [21]. Ostial splitting [60], ostium sphincteroplasty [58] and surgical angioplasty [17] can be also used to surgically correct the ALMCA. Few data exist regarding the long-term patency of the coronary artery and relief of symptoms of cardiac ischemia.

4.2. Percutaneous coronary angioplasty

Stents have considerable structural rigidity which may allow them to correct the ostial slit-like lumen and prevent the presumed interarterial compression during exercise. Intraprocedural use of transesophageal echocardiography is helpful in ascertaining that the stent was deployed in the correct position [24]. It is unclear whether this therapy, with its attendant risk of decreased long-term patency and the potential for ostial stenosis at the origin of the stent, is a durable solution. This therapy may be appropriate for an older patient with medical problems that would increase the risk of surgery or if they have co-existent atherosclerotic CAD that otherwise would be best treated with angioplasty and stent therapy [1].

4.3. Recommendations

Patients may be suspected to have ALMCA with an interarterial course, if they present with angina, abnormal non-invasive tests, myocardial infarction or cardiac arrest in an otherwise young healthy person or may be discovered serendipitously during investigation *e.g.* cardiac catheter for other pathology as CAD. Those patients should preferably be managed in the following manner:

All patients should undergo one or more of the novel non-invasive imaging techniques *e.g.* computerized assisted tomography, magnetic resonance imaging or transesophageal echocardiography that define clearly the origin and course of the ALMCA. For children and young adolescents, transthoracic echocardiography with color Doppler mapping is advised. Cardiac catheterization should be reserved for patients in whom the diagnosis is in question or in adult patients with risk factors for CAD. If catheterization is necessary, the best method of determining the origin and course of the anomalous coronary artery is to simultaneously inject in the orifice of the coronary artery and the pulmonary artery [30].

In symptomatic patients with ALMCA, indication for operative repair is irrefutable. The management of the asymptomatic patient remains somewhat uncertain but we are inclined towards surgical intervention to prevent the risk of sudden death. If surgical repair is not an option, avoidance of strenuous physical activity and competitive athletics should be urged. In symptomatic patients, repair should not be delayed. In the asymptomatic patient, we recommend delaying elective surgical repair until late puberty or approximately 10 years.

In elderly patients with co-existent CAD, coronary artery bypass graft seems to be the optimal therapy. If the origin of the ALMCA is at a level higher or cephalad to the commissure, unroofing technique is preferred. If the coronary artery is large, reimplantation is an attractive option. Close monitoring and long-term surveillance is critical post-procedure to detect ostial stenosis or AI.

For older patients with medical problems or co-existent atherosclerotic CAD, intracoronary stenting may be an alternative to surgical therapy, especially in emergency situations but the results need to be validated.

There is some limitation to the conclusions that can be drawn from the results of this review, mainly due to the lack of randomized controlled or large cohort studies in the literature. It is encouraging that this potentially lethal defect, can be repaired with low surgical risk, but the durability of these repairs is yet to be determined.

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