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Anomalous Left Coronary Artery from the Pulmonary Artery (ALCAPA): a Systematic Review and Historical Perspective

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Abstract

Purpose of the Review To provide an updated literature review for the diagnosis, treatment, and outcomes of anomalous left coronary artery from the pulmonary artery (ALCAPA).

Recent Findings The diagnosis of ALCAPA has shifted away from coronary angiography to noninvasive imaging modalities. Newer imaging techniques such as speckle tracking echocardiography to evaluate myocardial strain demonstrate subclinical myocardial dysfunction years after restoration of a dual coronary system.

Summary The diagnosis of ALCAPA is primarily with echocardiography and computed tomography coronary angiography. Coronary reimplantation is the preferred surgical technique. Long-term outcomes are excellent, greater than 97% survival in the modern era, and most patients will have resolution of their systolic dysfunction, with the most common indication for reintervention being mitral valve regurgitation. Some patients with successful reestablishment of a dual coronary arterial system have subclinical myocardial dysfunction, detected by myocardial strain echocardiography, in the setting of normal systolic and diastolic parameters. Further research is needed to determine the impact and long-term outcomes of subclinical dysfunction on long-term survivors.

Keywords Congenital heart disease · Anomalous coronary artery from the pulmonary artery · Coronary arteries · Myocardial strain imaging · Echocardiography

Introduction

Anomalous left coronary artery from the pulmonary artery (ALCAPA), also known as Bland-White-Garland syndrome, is a rare congenital heart defect that occurs in one in 300,000 live births [1•, 2–5, 6••, 7•, 8]. ALCAPA is one of the most common causes of myocardial ischemia in pediatrics, and early recognition and prompt surgical repair to reestablish a two coronary artery system is essential. With improvements in imaging technology, surgical advancements, and postoperative care, the outcomes for those diagnosed with ALCAPA are now excellent, for what was historically a near universally fatal disease.

The first description of anomalous coronary artery from the pulmonary artery was in 1866 by John Brooks, though his postmortem anatomical description was of an anomalous right coronary artery off the pulmonary artery [3]. In 1933, three physicians from Massachusetts General Hospital, William Bland, Paul White, and Joseph Garland, published the first clinical description of ALCAPA in a three-month-old boy with cardiomegaly, ischemic ECG changes, and paroxysmal episodes of distress precipitated by feeding [2, 3]. Dr. William Mustard attempted the first surgical correction of ALCAPA in 1953 performing a left carotid to ALCAPA end-to-end anastomosis [9]. Since then, multiple surgical techniques have been described for the repair, with the current procedure of choice being aortic reimplantation of the ALCAPA [2].

An updated classification scheme was developed as part of the Society for Thoracic Surgeons Congenital Heart Surgery

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Nomenclature and Database Project to include all possible anomalous pulmonary origins of coronary arteries (Table 1) [10]. Most commonly, the anomalous left coronary artery (LCA) arises from the left posterior-facing sinus of the pulmonary artery.

Pathophysiology

Coronary perfusion pressure is equal to the diastolic blood pressure in the great vessel minus the left ventricular end-diastolic pressure and is the driving force for perfusion of the myocardium. In utero, the coronary perfusion pressure in a fetus with ALCAPA is normal as the pulmonary artery pressure is high and similar to the aortic pressure. Immediately following birth, the pulmonary artery provides desaturated blood to the LCA, while the right coronary artery (RCA) is perfused normally with oxygenated blood from the aorta [11, 12••].

Over the coming days to weeks, as the pulmonary vascular resistance continues to fall, the diastolic blood pressure in the pulmonary artery falls, resulting in inadequate coronary perfusion pressure, reversal of flow in the LCA into the pulmonary artery, and resultant poor perfusion of the anterolateral left ventricular free wall and anterolateral papillary muscle. Collaterals from the RCA form postnatally and, depending on the degree and extent of coronary collateralization, the collateral flow can provide some myocardial perfusion. However, further pulmonary-coronary artery steal and the potential for myocardial ischemia occurs with a left-to-right shunt of blood from the RCA, through the collaterals, to the LCA, which preferentially shunts to the low-pressure pulmonary artery as opposed to flowing into the myocardium [11, 12••].

In the infantile form of ALCAPA, patients present with evidence of myocardial ischemia due to inadequate coronary collateralization and pulmonary-coronary artery steal. There is a subset of patients, however, who present in late childhood or adulthood with extensive coronary collateralization that provides adequate myocardial perfusion. Some of these patients may have had elevated pulmonary artery pressures or a restrictive LCA orifice into the pulmonary artery that alters the pathophysiology such that they may reach adulthood with minimal symptoms [11, 12••, 13].

Table 1 Classification scheme from the Society for Thoracic Surgeons Congenital Heart Surgery Nomenclature and Database Project, adapted from Dodge-Khatmai et al. [10]

Classification of anomalous pulmonary origins of coronary arteries
1) Anomalous origin of the left main coronary artery from the pulmonary artery (most common)
2) Anomalous origin of the right coronary artery from the pulmonary artery
3) Anomalous origin of the circumflex coronary artery from pulmonary artery
4) Anomalous right and left coronary arteries from the pulmonary artery

Clinical Features

For patients with the infantile form of ALCAPA, with little or no collaterals, there is an early onset of symptoms, classically at 1–2 months of age, accompanied by progressive myocardial ischemia, left ventricular dysfunction, and mitral regurgitation that can result in sudden death [12••]. Caregivers may describe signs of angina precipitated by feeding with crying, tachypnea, and diaphoresis and decreased oral intake. Feeding intolerance can be mistaken for a gastrointestinal problem. On examination, infants may have evidence of failure to thrive, tachypnea, tachycardia, wheezing, grunting, diaphoresis, and hepatomegaly and can present in cardiogenic shock. Patients may have a holosystolic murmur from mitral insufficiency, a gallop, and possibly a soft continuous murmur appreciated best at the left-upper sternal border from the retrograde coronary flow into the pulmonary artery. Older children and adults who have extensive coronary collateralization can have a wide range of presentations, from asymptomatic, to a mitral insufficiency murmur, to syncope and even sudden death.

Electrocardiogram may demonstrate evidence of an anterolateral infarct pattern with pathologic Q waves in lead I, a VL and V4 through V6 and/or abnormal R wave progression in the precordial leads [14]. Chest radiograph may demonstrate an enlarged cardiac silhouette and pulmonary edema. Cardiac enzymes may or may not be elevated at time of presentation, depending on whether there is ongoing ischemia. Plasma B-type natriuretic peptide levels are typically elevated due to ventricular dysfunction.

Natural History

In the infantile form of ALCAPA, mortality has been reported to be > 80% without intervention [15, 16]. The adult-type of ALCAPA (10–15% of patients), [17] with significant collaterals, may be asymptomatic for decades due to the collateralization from the right coronary artery. A literature review of 151 adult patients with ALCAPA found that 14% were asymptomatic and 62% of those with life-threatening presentations were asymptomatic prior to diagnosis [13]. Among these patients, there is an 80–90% incidence of sudden cardiac death at 35 years, with lower risk of sudden cardiac death after age 50 [13, 18–22]. Given the high incidence of sudden cardiac death, surgical intervention is warranted once the diagnosis is made [12••].

Diagnosis

A high index of suspicion is critical, and a thorough evaluation of the origins and direction of flow in the coronary arteries is essential for any infant or child that presents with left ventricular dysfunction. The gold standard for diagnosis of ALCAPA is coronary artery angiography via cardiac catheterization with an aortogram that demonstrates an enlarged RCA with coronary collaterals and retrograde filling of the pulmonary artery from the LCA. In the modern era, the diagnosis can often be made using noninvasive imaging modalities, and cardiac catheterization is now more commonly reserved for uncertain cases [6••, 23•].

With advancements in imaging technology, the diagnostic accuracy of ALCAPA with echocardiography has improved over time [6••]. Two-dimensional (2-D) imaging reveals an enlarged RCA and the LCA arising anonymously from the pulmonary artery, though the LCA often courses very closely to the left aortic sinus of Valsalva and the diagnosis based on 2-D imaging alone can be easily missed. In addition to depressed left ventricular systolic function, the anterolateral papillary muscle and left ventricular endocardium may have increased echogenicity, indicative of ischemia and varying degrees of mitral insufficiency are invariably present. Color Doppler echocardiography can demonstrate retrograde coronary blood flow in the LCA entering into the pulmonary artery. In a recent analysis of diagnostic echocardiographic features of ALCAPA, Patel et al. described the most common findings that are highly suggestive of ALCAPA in infants and children, including LCA flow reversal (91%), visualization of collateral coronary arteries (85%), dilated RCA for age (81%), retrograde flow into the pulmonary artery (79%), moderate to severe mitral valve regurgitation (74%), left ventricular dysfunction (66%), and endocardial fibroelastosis (57%) [6••]. A majority of echocardiographic studies in patients with ALCAPA will have at least five of these ancillary markers present [6••].

Given the vastly different management strategies for cardiac dysfunction secondary to dilated cardiomyopathy or myocarditis versus ALCAPA, additional imaging modalities can aid in establishing the diagnosis. At our center, those with suspected ALCAPA on echocardiography undergo confirmatory computed tomography (CT) coronary angiography, which allows for excellent spatial resolution to delineate the origin and course of the coronary arteries. Cardiac magnetic resonance imaging (MRI) may not provide the spatial resolution that cardiac CT does, though it offers the added benefit of late gadolinium enhancement which can be indicative of cardiac fibrosis secondary to chronic ischemia. Both CT and MRI techniques allow for three-dimensional reconstructed images, which can be useful in preoperative surgical planning [24•, 25]. Transesophageal echocardiography can also aid in the diagnosis, though is typically performed intraoperatively to confirm the anatomical findings and evaluate left ventricular function and degree of mitral regurgitation immediately prior to surgical repair.

Newer echocardiographic techniques such as myocardial strain (speckle tracking) and nuclear myocardial perfusion imaging may offer additional insight to cardiac dysfunction, though they are not specific in making the diagnosis and are likely more beneficial in follow-up after surgical repair, as described below [1•].

Treatment

Surgical Repair

Surgical intervention is indicated for all patients diagnosed with ALCAPA. Medical treatment has demonstrated poor survival (45–100%) mortality [17, 19, 26] and plays no role in the current era [12••]. Current recommendations are surgical intervention at the time of diagnosis, with surgery performed within days of diagnosis given the importance of early intervention [11]. Early surgical intervention is indicated even in adults with apparent adequate collateralization given these patients are at risk for sudden cardiac death [27••].

Surgical strategies for this defect have evolved over time. One of the earliest palliative interventions was reported by Dr. Willis Potts, who utilized an aortopulmonary shunt to increase pulmonary pressure and oxygenated blood flow to the anomalous coronary artery [28]. In 1953, Mustard reported attempting a left common carotid artery to ALCAPA end-to-end anastomosis for this lesion [29]. The first successful surgical intervention was reported by Sabiston, who ligated the ALCAPA at its origin, to prevent coronary steal into the pulmonary artery [30]. Other strategies have included saphenous vein graft to ALCAPA as reported by Cooley in 1966 [31] and left subclavian artery to ALCAPA bypass reported by Meyer in 1968 [32].

The current surgical management of ALCAPA is reimplantation of the left coronary directly onto the aorta as a coronary button, as first described by Neches et al. in 1974 (Figs. 1 and 2) [33, 34]. Operative outcomes of ALCAPA are now excellent, with recent series describing no perioperative mortality or early mortality [2, 6••]. This technique can be applied regardless of the size of patient or type of ALCAPA; however, depending on the location of the origin of the ALCAPA, direct reimplantation is not always technically feasible requiring alternate methods [34].

For patients with inadequate coronary length or unfavorable anatomy for reimplantation, Takeuchi described creating a baffle through the pulmonary artery lumen into the aorta along with a pericardial patch of the anterior pulmonary artery to prevent pulmonary artery stenosis [35]. This technique allows reestablishment of a two coronary system regardless of

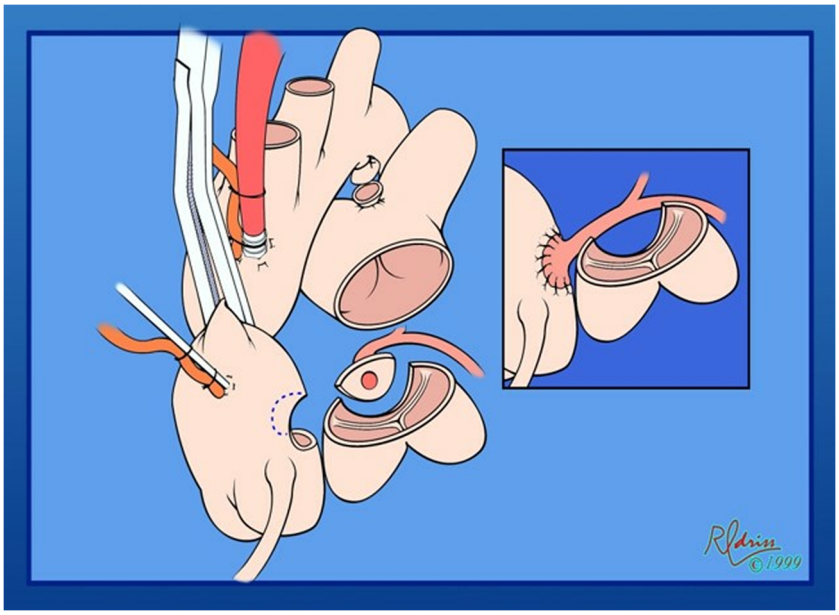

Fig. 1 After the second dose of cardioplegia, an opening is created in the left posterolateral wall of the ascending aorta for implantation of the anomalous left coronary button. Care is taken not to injure the aortic valve. This opening is typically approximately one-third smaller in size than the button that was created. The large button of coronary artery can then act as a "conduit" for elongation of the left coronary artery. With proper mobilization of the left coronary artery, it is usually quite easy to perform this anastomosis. Once the anastomosis is created (inset), the aortic cross-clamp is removed, and now both right and left coronary arteries are directly perfused. (Reproduced with permission from Backer CL, Hillman N, Dodge-Khatami A, Mavroudis C: Anomalous origin of the left coronary artery from the pulmonary artery: successful surgical strategy without assist devices. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu 2000;3:165–72)

the distance of the ALCAPA from the aorta with long-term survival of 78–100%, comparable with other techniques [36]. However, a high right coronary artery origin or ALCAPA origin adjacent to pulmonary cusp can complicate this repair technique [37]. Complications specifically related to the Takeuchi procedure include baffle leak creating a coronary-

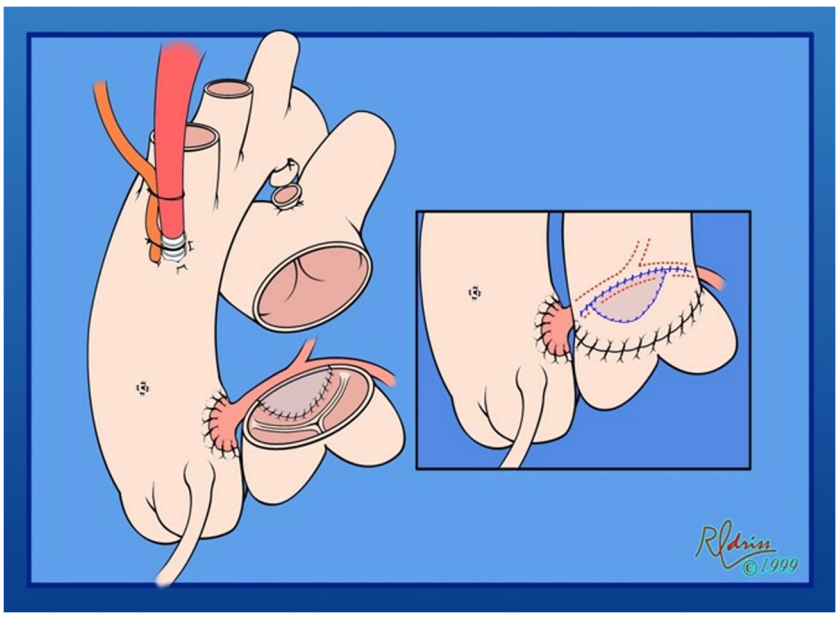

Fig. 2 The aortic cross-clamp is off. The posterior sinus of the pulmonary artery where the button was harvested is reconstructed with a patch of fresh autologous pericardium. The pulmonary artery is reanastomosed at the site of the transection (inset). This reconstruction of the pulmonary artery with the cross-clamp off helps to minimize the aortic cross-clamp time. In almost all instances, it is possible to perform the entire procedure with two doses of cardioplegia given in the sequence described. (Reproduced with permission from Backer CL, Hillman N, Dodge-Khatami A, Mavroudis C: Anomalous origin of the left coronary artery from the pulmonary artery: successful surgical strategy without assist devices. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu 2000;3:165–72)

pulmonary artery fistula (27%), supravalvar pulmonary stenosis (24%), and aortic valve insufficiency [12••, 38].The reoperation or reintervention rate for the Takeuchi method has been reported to be as high as 30%, with an average across studies in the literature of 23% [18, 39, 40].

Other techniques have been described to manage inadequate coronary length, such as free subclavian artery interposition [19]. This technique can be performed in young patients, allows for growth with long-term patency of 60–80% [41, 42], and has an operative mortality of 0–29% [32, 41–43]. However, concerns have been reported regarding the risk of the artery kinking at its origin from the aorta and inadequate length [34, 42]. Saphenous bypass grafting, while technically suitable in older children and young adults, has a reported mortality of 0–38%, with a high potential for late graft occlusion, making the technique less than ideal [19, 44]. In adults with ALCAPA, direct reimplantation can be challenging due to the decreased mobility, calcification, and friability of tissues [11, 17, 20]. In some of these patients, left internal mammary bypass grafting with ligation of the origin of the ALCAPA may need to be considered [45, 46].

Irrespective of the technique employed, management of cardioplegia administration for adequate myocardial protection during the operation is critical. The most common method is to inject cardioplegia into the aortic root while occluding the main pulmonary artery or pulmonary arteries individually or occluding the ALCAPA at the pulmonary origin [34, 40, 47, 48]. Others will inject cardioplegia directly into the pulmonary trunk in addition to aortic administration of cardioplegia [48, 49].

Mitral valve repair at the time of initial surgical repair for ALCAPA remains controversial. A majority of surgeons, including those at our center, do not advocate this approach at least during the initial surgery [11, 19, 34, 39, 47, 49, 50]. Most patients will have improvement in mitral valve regurgitation once normal coronary flow is reestablished without surgical interventions on the mitral valve itself [2, 4, 6, 8]. This is particularly true for patients who are less than 1 year of age [51•]. In fact, lack of improvement in mitral regurgitation or worsening mitral regurgitation postoperatively may indicate inadequacy or stenosis of the ALCAPA repair [11, 12••, 50]. Some authors suggest that the longer cross clamp time for mitral valve repair may be harmful and does not affect the postsurgical outcome or ventricular function [2].

Surgical correction of ALCAPA reestablishing a two coronary system, whether it be via direct reimplantation or baffle, also corrects sequelae related to ALCAPA, including mitral regurgitation and left ventricular dysfunction and dilation if the myocardium remains viable [11, 12••, 18, 39, 47, 49, 52, 53]. There is no evidence demonstrating superiority of a single technique of surgical restoration of a two coronary system in terms of long-term left ventricular function or late mortality, but there is reduced survival seen with ALCAPA ligation, in which normalization of left ventricular volume and ejection fraction does not occur [11, 18, 36, 39, 52, 53]. Scarred, nonviable myocardium will not demonstrate recovery, but resection of aneurysmal, scarred ventricular tissue is rarely justified [11, 47, 48].

Mechanical Support

Support after cardiopulmonary bypass may be necessary, particularly in those with poor ventricular function preoperatively, patients with stunned myocardium or persistent arrhythmias. Previously, some critically ill infants were considered too-high risk for reimplantation with some advocating for ligation of the ALCAPA, [54], which has been associated with poor outcomes [18, 19, 34, 55–57]. With improvements in mechanical circulatory support, if necessary, patients can be supported with extracorporeal membrane oxygenation or left ventricular assist device (LVAD) as a bridge to recovery [21, 39, 50, 58, 59, 60••]. In the more recent era, the Berlin Heart EXCOR (EXCOR Pediatric, Berlin Heart Inc., The Woodlands, Texas) or continuous-flow LVADs have been used for temporary support with good outcomes for those who fail to wean from cardiopulmonary bypass [58, 59]. Predictors of patients who will require LVAD support postoperatively include severe preoperative dysfunction and prolonged aortic cross clamp time [58].

Outcomes and Follow-Up

Overall outcomes remain excellent with greater than 85% long-term survival [2, 4, 36, 51•, 60••, 61•] in all patients, and greater than 95% survival at 20 years in patients repaired in infancy [2, 25, 61•]. There is an era effect with a recent report from Germany describing 97% of patients surviving up to 20 years if repaired after 1995 [2]. Regardless of age at the time of operation, reestablishment of a two coronary artery system results in recovery of left ventricular systolic function in 75–90% of patients, even in patients who had severe left ventricular systolic dysfunction preoperatively [2, 4, 6, 51•, 60••, 61•]. Many patients will have some degree of mitral valve regurgitation postoperatively, but a small percentage of patients who did not undergo concomitant mitral valve repair (3–14%) require postoperative intervention on the mitral valve [2, 61•]. Stenosis of the reimplanted coronary artery remains a rare complication [60••, 61•]. Over time, the RCA will regress to a normal size and regression of right-sided collateralization has been observed [11]. Freedom from any reoperation remains excellent at 88% at 5 and 10 years [6••] and 76% at 20 years after the initial operation [4, 61•].

Following ALCAPA repair, patients require lifelong follow-up. Median time to normalization of global left ventricular systolic function has been reported at 91 days (range 5–

429 days) in a group of 25 patients for which long-term echocardiography data was available [62••]. Once discharged from the intensive care unit, many patients are treated with an oral heart failure regimen, with one report noted that diuretics, digoxin, ACE inhibitors, and/or aspirin were common medications at discharge [62••].

While improvement in global left ventricular systolic function is generally the rule, newer echocardiographic techniques are beginning to give further insight into long-term functional assessment. In postoperative ALCAPA patients with normalized systolic function, impairments have been noted in measures of diastolic dysfunction and myocardial strain imaging [1•, 63]. Using speckle tracking echocardiography, Castaldi et al. showed the longitudinal and circumferential strain was reduced in the subendocardial regions of the left coronary artery compared with subendocardial regions of the right coronary artery in normal subjects, while radial strain was preserved [1•, 60••, 64•, 65]. Confirmatory cardiac MRI was performed in the subset of patients with abnormal strain and demonstrated either left coronary artery stenosis or late gadolinium enhancement indicating fibrosis in the regions of abnormal myocardial strain. These abnormalities can be seen even in patients who have a normal cardiopulmonary exercise stress test [1•].

Adult congenital heart disease guidelines, from the American College of Cardiology and American Heart Association, recommend that patients with prior history of surgical repair for ALCAPA undergo noninvasive stress imaging every 3–5 years for routine surveillance [66]. Stress echocardiography, single-photon emission CT, and stress MRI can be used to assess myocardial ischemia following repair [67]. Schmitt et al. analyzed 21 patients with a median of 10-year follow-up and MRI at rest demonstrated myocardial scars and regional wall motion abnormalities in 67% and perfusion deficits in 28%. Dobutamine stress MRI, however, detected an additional 19% of patients with wall motion abnormalities and 14% with perfusion deficits not observed at rest [67]. The long-term implications of these abnormalities are not yet well understood, and routine surveillance with echocardiography, Holter monitors, cardiopulmonary exercise testing, and stress imaging is recommended.

Conclusions

While ALCAPA is a rare congenital heart defect, early and accurate diagnosis is essential to management and long-term survival. LCA coronary reimplantation to the aorta is the preferred surgical technique when feasible. Reestablishment of two coronary artery supply typically results in normalization of left ventricular function at follow-up. In this era of excellent surgical outcomes and improved perioperative management, we must shift our attention to better recognition and monitoring of subtler impairments in left ventricular function with close follow-up of the long-term survivor of ALCAPA.

Compliance with Ethical Standards

Conflict of Interest Gary S. Beasley, Elizabeth H. Stephens, and Anna Joong declare no conflict of interest.

Carl L. Backer received consultant fees through W.L. Gore and Associates.

Human and Animal Rights and Informed Consent This article does not contain any studies with human or animal subjects performed by any of the authors.

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