International Journal of Cardiology and Cardiovascular Diseases

Miller L, et al., Int J Cardiol Cardiovasc Dis. 2026;6(1):24-29.

Short Communication

Spectrum of congenital coronary artery anomalies in hypertrophic cardiomyopathy: a systematic review

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Received date: November 28, 2025
Accepted date: March 13, 2026

Abstract

Myocardial ischemia is a frequent finding in hypertrophic cardiomyopathy (HCM) potentially resulting from microvascular dysfunction, extravascular compression, reduced coronary flow reserve, hemodynamic abnormalities, or concomitant atherosclerotic coronary artery disease (CAD). From June 1991 to October 2024, 40 congenital coronary artery anomalies (CCAA) were reported in 30 adults with HCM [age 18–87 (47.6 ± 19.7); 60% male; 37% obstructive; 20% apical variant]. Clinical data on ischemia, therapy, and outcomes were reviewed. Coronary anatomy was categorized by number of ostia: single (n=12; 40%), two (n=13; 43%), three (n=4; 13%), or four (n=1; 4%). The anomalous vessel was the left main (20%), left anterior descending (25%), left circumflex (18%), right coronary artery (35%), or ramus intermedius (2%). Two patients (7%) with apical HCM also had coronary fistulae. Myocardial ischemia was present in 13 patients (43%), including 5 with anomalous vessels coursing between the great vessels. Two of five patients with sudden cardiac arrest had a high-risk anomaly. Pharmacologic therapy was provided to 13 (43%), coronary surgery to 3 (10%), and septal reduction to 8 (27%). Among 15 patients with follow-up, all reported symptomatic improvement. CCAA may occur in HCM and contribute to ischemia; management should be guided by anomaly type and objective evidence of ischemia.

Keywords: Hypertrophic cardiomyopathy, HCM, Coronary artery, Coronary anomalies, Myocardial ischemia

Abbreviations: CAD: Coronary Artery Disease; CCAA: Congenital Coronary Artery Anomalies HCM: Hypertrophic Cardiomyopathy SCA: Sudden Cardiac Arrest

Introduction

Myocardial ischemia is frequently detected in adults with hypertrophic cardiomyopathy (HCM) [1] and is commonly attributed to microvascular disease, extravascular compressive forces, reduced coronary flow reserve, hemodynamic abnormalities, and concomitant atherosclerotic coronary artery disease (CAD) [2]. Congenital coronary artery anomalies (CCAA) may be present in some adults with HCM with potential independent contribution to myocardial ischemia. However, such association has not been systematically evaluated. We aimed to examine the spectrum of reported CCAA in adults with HCM and their potential association with myocardial ischemia.

Methods

A systematic, worldwide literature search, using keywords of "hypertrophic cardiomyopathy and congenital coronary anomalies", was conducted across three databases: PubMed/MEDLINE, Google Scholar, and the Temple University Library search engine to identify published case reports and case series highlighting patients with both HCM and CCAA (**Figure 1**). In addition to database searching, reference lists of all retrieved articles were manually screened to identify additional eligible studies not

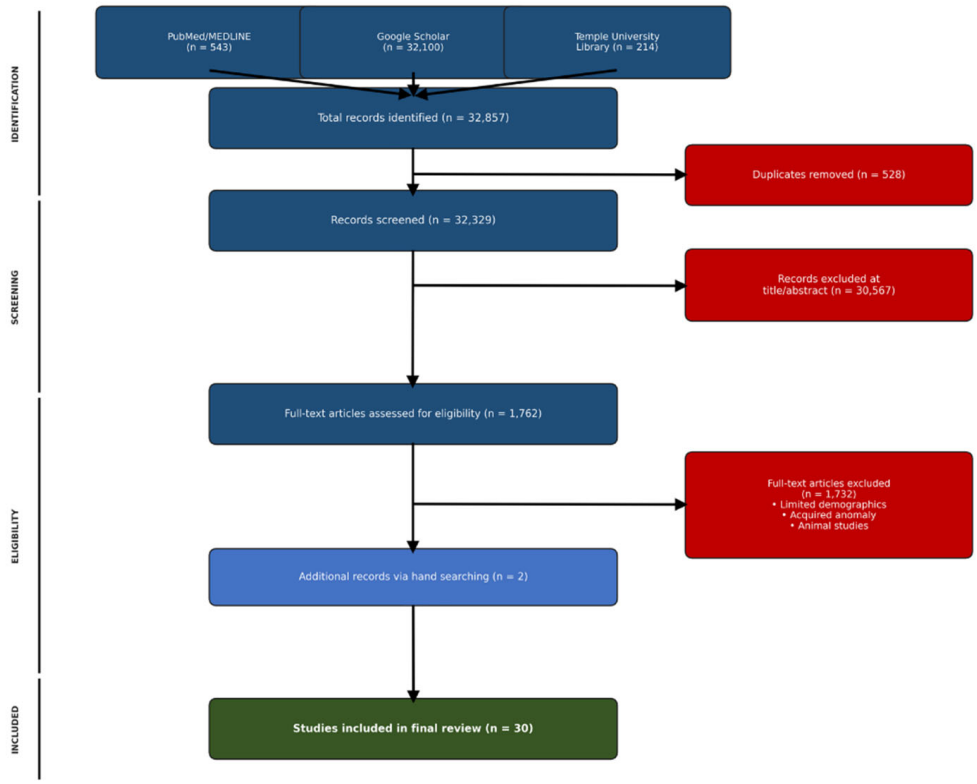

Figure 1. PRISMA flow diagram illustrating the systematic search and study selection.

captured through the primary search. Studies were included if they met all of the following criteria: (1) reported at least one patient; (2) included a diagnosis of HCM as reported by the original authors; (3) described the presence of one or more congenital coronary artery anomalies; and (4) provided sufficient patient-level data to identify both patient demographics (age and sex) and the type and identity of the anomalous coronary vessel and its anatomy. Case reports and case series were eligible. No language restrictions were applied.

Exclusion criteria were as follows: (1) animal studies; (2) cases in which the coronary anomaly was acquired rather than congenital in etiology (e.g., secondary to Kawasaki disease or atherosclerosis); (3) cases lacking patient demographic data (age or sex) or an adequate description of the coronary anomaly; and (4) duplicate reports identified as describing the same patient across multiple publications, in which case only the most detailed or original report was retained.

Clinical data on patient history, family history, the anomalous vessel(s), presence or absence of myocardial ischemia, mode of therapy, and outcomes were extracted from each case. A subset of these characteristics can be found in **Table 1**. From June 1991 to October 2024, a total of 40 individual anomalous arteries were reported in 30 adults with HCM. Institutional review board approval was not required as all data were obtained from de-identified, previously published case reports and case series.

Results

Patients were 18 to 87 (47.6 ±19.7) years old at the time of report; 60% were male; 37% had an obstructive phenotype of HCM while 20% demonstrated the apical variant. The coronary anatomy of the individual cases were graphically reproduced (**Figure 2**) and were categorized based on the number of coronary ostia present in the aorta {single ostium [n=12 (40%)]; 2 ostia [n=13 (43%)]; 3 ostia [n=4 (13%)] and 4 ostia [n=1 (4%)}. The anomalous coronary artery was the left main coronary artery in 8 (20%); left anterior descending coronary artery in 10 (25%); left circumflex coronary artery in 7 (18%); right coronary artery in 14 (35%) or the ramus intermedius coronary artery in 1 (2%). Two patients (7%) with apical variant HCM also had coronary artery fistulae in the left anterior descending coronary artery. Overall, there were 38 instances of anomalous origin, 33 instances of anomalous course and 2 instances of anomalous termination (fistulae) among the congenitally abnormal coronary arteries. Of the 30 patients reviewed, 13 (43%) showed objective evidence of myocardial ischemia with 5 (38%) having an anomalous coronary artery coursing in between the great arteries. In addition, 2 of 5 patients with sudden cardiac death had a high-risk coronary anomaly, defined as either having a slit-like ostium, acute take-off angle or coursing in between the great arteries. Pharmacologic treatment was offered to 13 (43%) patients, and 3 (10%) patients underwent coronary artery surgery (bypass grafting in 2 and unroofing an intramural epicardial coronary artery in 1). Septal reduction therapy was performed in 8 (27%). Follow up data was available in 15 patients including 2 of the 3 who underwent surgical correction. Symptomatic improvement was noted on follow up in all 15.

Table 1. Bibliographical information for cases with HCM and CCAA.

Figure #1 Reference	Author [manuscript reference]	Publication Year	Journal	Age	HCM Phenotype	Presence of Ischemia?
XVII	Liddy [20]	2014	BMJ	40	HCM	No
XXIII	Moza [26]	2011	AMJS	29	HOCM	NA
XVIII	Tyczynski [21]	2013	Studium Przypadku	65	HOCM	Yes, dipyridamole induced
VII	Atmaca [10]	2002	Journal of Invasive Cardiology	38	HCM	No
XIII	Niwayama [16]	1991	Kokyu To Junkan	47	HCM	Yes, exercise stress induced
VIII	Lv [11]	2022	Journal of Cardiothoracic Surgery	64	HOCM	NA
XIV	Georgekutty [17]	2014	Cardiology Young	18	Apical HCM	Yes
I	Chow [4]	2009	The Canadian Journal of Cardio.	70	Apical HCM	Yes, dipyridamole induced
XV	Alqarqaz [18	2011	Journal of Cardiovascular Medicine	44	HCM	Yes, SPECT positive
XXVI	Ferreira [29]	2008	International Journal of Cardiology	26	HCM	NA
XXVIII	Georgiadou [31]	2006	International Journal of Cardiology	68	HCM	Yes, elevated troponins
XXI	Dermengiu [24]	2010	Romanian Journal of Legal Medicine	20	HCM	NA
XXX	Beach [33]	2001	Arch Pathol Lab Med	49	HCM	NA
XXVIII	Bush [30]	2005	Images in Cardiovascular Medicine	87	HOCM	NA
XXIX	Cifcti [32]	2009	Cardiology in the Young	31	HCM	Yes, exercise stress induced
IX	Zuccarino [12	2009	European Soceity of Cardiology	23	HCM	Yes, elevated troponins
X	Kurflaklıoğlu [13]	2005	Olgu Sunumu	22	HOCM	NA
III	Serino [6]	1998	Heart	71	HOCM	NA
IV	Hara [7]	2006	Journal of Invasive Cardiology	80	HOCM	NA
XI	Efthimiadis [14]	2013	Herz	45	HCM	No
V	Muller [8]	2023	JACC	77	Apical HCM	Yes, elevated troponins
XX	Santos [23]	2017	JACC	57	Apical HCM	Yes, dobutamine stress induced
XIX	Sarkar [22]	2019	CHEST	66	Apical HCM	Yes, exercise stress induced
VI	Penciu [9]	2014	Ann Thoracic Surgery	48	HOCM	Yes
XXIV	Sato [27]	2016	HEARTS Original	41	HCM	NA
XXII	Zheng [25]	2019	BMC Cardiovascular Disorders	26	HCM	NA
XII	Sinha [15]	2022	ARYA Athero	21	HOCM	No
XVI	Guo [19]	2015	Angiology	55	Apical HCM	No
XXV	Woznica [28]	2018	Kardiologia Polska	61	HOCM	NA
II	O'Gorman [5]	2024	JACC Case Report	39	HOCM	Yes, elevated troponins

NA: Not Available; HCM: Non-obstructive Hypertrophic Cardiomyopathy; HOCM: Obstructive Hypertrophic Cardiomyopathy; SPECT: Single Photon Emission Computerized Tomography

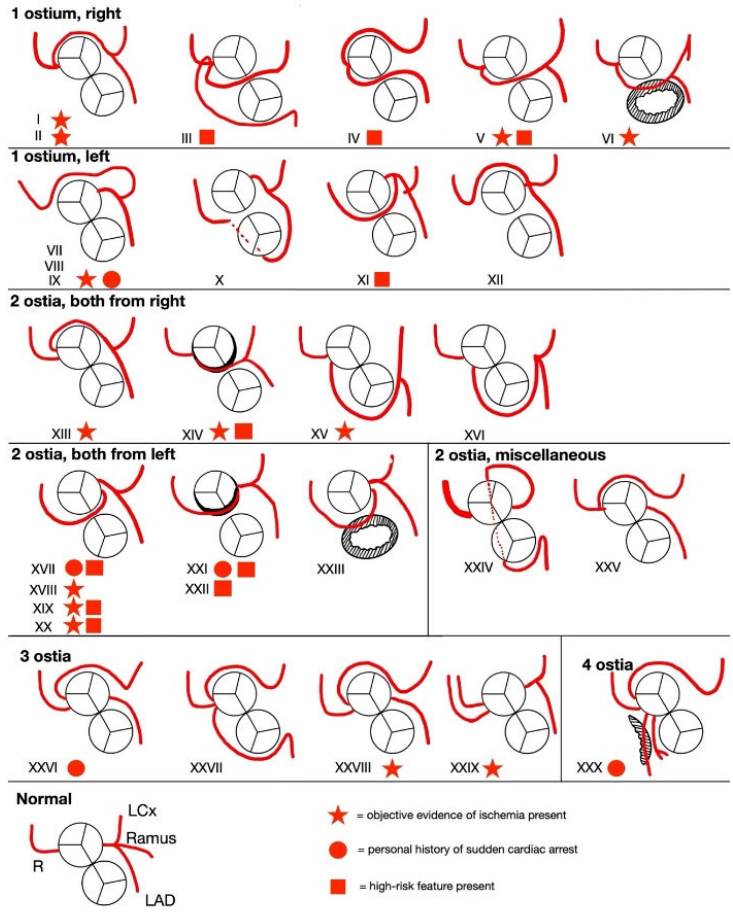

Figure 2. Graphical representation of origin and course of coronary artery anomalies.

Discussion

CCAA refers to a heterogeneous group of coronary malformations divided into three main categories: anomalies of origin, anomalies of course, and anomalies of termination [3]. The literature of each main CCAA category within HCM patients is limited; however, one of the most studied sub-variants are myocardial bridges which are nearly twice as prevalent in HCM patients and are theorized to potentially contribute to myocardial ischemia and SCD [4]. Beyond myocardial bridges, there is sparse literature which describes anomalies of origin and anomalies of termination in HCM patients.

Our study shows that CCAA may be concomitantly present in patients with phenotypic HCM and in certain cases (~43%) may be associated with myocardial ischemia. Of interest, most reported cases (~63%) had the non-obstructive phenotype and the 2 patients with coronary cameral fistula had the apical variant of HCM [34]. While the right coronary was involved in ~35% of these anomalies, the remaining cases were similarly distributed among left main (8), left descending (10) and left circumflex (7) coronary arteries. Overall, coronary anatomy varied widely among patients with HCM and no predominant specific anomaly was identifiable. There were no instances of tunneled (bridged) coronary artery among patients with anomalous coronary artery.

Among the 30 cases, 12 (40%) were considered to have the "high risk" features of coursing in between the great vessels, having a slit-like ostium, or having an acute take-off angle. Ischemia was present in 5 of the latter 12 patients. Only 2 of 5 patients with sudden cardiac arrest (SCA) had a "high risk" coronary anomaly. Further research into the risk of SCD in patients with HCM and CCAAs will help optimize treatment of this population.

While it can be speculated that the presence of CCAA would heighten risk of sudden cardiac death among competitive athletes, only one of the cases reported died suddenly during soccer training session. Based on our observations from these published case reports, patients with CCAA and HCM present in three main categories: asymptomatic and subsequently found incidentally, symptomatic with recurrent angina or syncope on exertion found via echocardiogram or coronary angiography, and post-mortem autopsies secondary to SCA.

Conclusion

The true prevalence of CCAA in patients with HCM is not known. CCAA is rarely acknowledged as a potential contributor to myocardial ischemia in the presence of multiple other potential etiologies (microvascular dysfunction, extravascular compressive

forces, reduced coronary flow reserve, hemodynamic abnormalities, intramyocardial course, and concomitant atherosclerotic coronary artery disease) in patients with HCM. While evidence-based treatment strategies are lacking for specific management of coronary anomalies in patients with HCM, application of general approaches to CCAA appears to have been effective in most cases. Further studies are needed to understand the true scope of CCAA in HCM and its impact on treatment and outcomes of the patients.

Conflicts of Interest

The authors declare that we have no conflicts of interest.

Funding

Not applicable.

Acknowledgements

Not applicable.

Author Contributions

LM: Conceptualization, Data curation, Formal analysis, Investigation, Visualization, Writing-original draft, Writing-review & editing. MS, KG, NR: Writing-original draft, Writing-review and editing. JS: Conceptualization, Formal analysis, Supervision, Validation, Writing-original draft, Project administration, Writing-review & editing. All authors read and approved the submitted version.

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