

Supplemental Information

SUPPLEMENTAL TABLE 3 Full Electronic Search Strategy From Ovid Medline

Number	Searches	Results
1	Mucocutaneous Lymph Node Syndrome/	5992
2	(Kawasaki Disease or Kawasaki syndrome or mucocutaneous lymph node syndrome or viral mucocutaneous lymphadenopathy).tw,kf.	6841
3	or/1-2	7482
4	exp Mortality/	380 099
5	(death rate? or mortalit*).tw,kf.	777 333
6	exp Myocardial Infarction/	174 531
7	(cardiovascular stroke? or heart attack? or myocardial infarct*).tw,kf.	202 596
8	exp Heart Failure/	120 817
9	(cardiac failure? or heart decompensation or heart failure?).tw,kf.	182 888
10	exp Hypertension/	253 371
11	(high blood pressure? or hypertension).tw,kf.	397 128
12	Coronary Artery Disease/	61 642
13	(coronary arteriosclerosis#s or coronary artery disease? or coronary atheroscleroses or coronary atherosclerosis).tw,kf.	93 806
14	exp Stroke/	133 595
15	(stroke? or apoplexy or brain vascular accident? or cva or cvas or cerebrovascular accident? or cerebrovascular apoplexy).tw,kf.	259 577
16	Cerebral Revascularization/	3804
17	exp Myocardial Revascularization/	91 611
18	(revascularization? or "ec ic arterial bypass*" or extracranial intracranial arterial bypass* or "sta mca bypass*" or internal mammary artery implantation).tw,kf.	52 589
19	Infections/	38 713
20	infection?.tw,kf.	1 409 792
21	exp Eczema/	11 170
22	eczema*.tw,kf.	19 794
23	exp Asthma/	127 486
24	asthma*.tw,kf.	158 950
25	exp Psoriasis/	39 875
26	(palmoplantar pustulosis or psoriasis#s or "pustular psoriasis of palms and soles" or "pustulosis palmaris et plantaris" or "pustulosis of palms and soles").tw,kf.	40 854
27	exp Arthritis/	258 187
28	(inflammat* adj3 arthriti*).tw,kf.	9559
29	or/4-28	3810 549
30	3 and 29	2241
31	randomized controlled trial.pt.	507 991
32	clinical trial.pt.	523 316
33	randomi?ed.ti,ab.	623 251
34	placebo.ti,ab.	214 373
35	dt.fs.	2 212 640
36	randomly.ti,ab.	336 337
37	trial.ti,ab.	595 071
38	groups.ti,ab.	2 084 947
39	or/31-38	4 914 292
40	Epidemiologic studies/	8337
41	exp case control studies/	1 084 693
42	exp cohort studies/	2 001 766
43	Case control.tw.	125 051

SUPPLEMENTAL TABLE 3 Continued

Number	Searches	Results
44	(cohort adj (study or studies)).tw.	205 141
45	Cohort analy\$.tw.	8020
46	(Follow up adj (study or studies)).tw.	49 145
47	(observational adj (study or studies)).tw.	106 318
48	Longitudinal.tw.	244 196
49	Retrospective.tw.	528 429
50	Cross sectional.tw.	351 177
51	Cross-sectional studies/	329 821
52	or/40-51	3 002 521
53	39 or 52	6 941 441
54	3 and 29 and 53	909
55	animals/	6 621 449
56	humans/	18 537 815
57	55 not (55 and 56)	4 675 883
58	54 not 57	905

Ovid MEDLINE(R) and Epub ahead of print, in process and other nonindexed citations and daily 1946 to June 19, 2020.

SUPPLEMENTAL TABLE 4 Full Electronic Search Strategy From Embase

Number	Searches	Results
1	Mucocutaneous Lymph Node Syndrome/	11 410
2	(kawasaki disease or kawasaki syndrome or mucocutaneous lymph node syndrome or viral mucocutaneous lymphadenopathy).tw,kw.	9492
3	or/1-2	11 903
4	exp mortality/	1 135 715
5	(death rate? or mortalit*).tw,kw.	1 178 432
6	exp heart infarction/	403 539
7	(cardiovascular stroke? or heart attack? or myocardial infarct*).tw,kw.	306 098
8	exp heart failure/	531 632
9	(cardiac failure? or heart decompensation or heart failure?).tw,kw.	311 863
10	exp hypertension/	772 945
11	(high blood pressure? or hypertension).tw,kw.	643 385
12	exp coronary artery disease/	340 425
13	(coronary arterioscleros#s or coronary artery disease? or coronary atheroscleroses or coronary atherosclerosis).tw,kw.	152 297
14	exp cerebrovascular accident/	213 364
15	(stroke? or apoplexy or brain vascular accident? or cva or cvs or cerebrovascular accident? or cerebrovascular apoplexy).tw,kw.	424 192
16	cerebral revascularization/	2527
17	heart muscle revascularization/	32 565
18	(revascularization? or "ec ic arterial bypass*" or extracranial intracranial arterial bypass* or "sta mca bypass*" or internal mammary artery implantation).tw,kw.	82834
19	exp infection/	3 742 248
20	infection?.tw,kw.	1 900 611
21	exp eczema/	33 726
22	eczema*.tw,kw.	34 631
23	exp asthma/	275 612
24	asthma*.tw,kw.	243 871
25	exp psoriasis/	93 781
26	(palmoplantaris pustulosis or psoriasis#s or "pustular psoriasis of palms and soles" or "pustulosis palmaris et plantaris" or "pustulosis of palms and soles").tw,kw.	65 599
27	exp arthritis/	511 380
28	(inflammat* adj3 arthriti*).tw,kw.	16 808
29	or/4-28	7 837 604
30	3 and 29	7548
31	exp clinical trial/	1 518 570
32	randomi?ed.ti,ab.	894 606
33	placebo.ti,ab.	311 892
34	dt.fs.	3 891 806
35	randomly.ti,ab.	447 350
36	trial.ti,ab.	872 660
37	groups.ti,ab.	2 984 539
38	or/31-37	7 764 706
39	Clinical study/	169 710
40	case control study/	156 729
41	Family study/	27 116
42	Longitudinal study/	141 354
43	Retrospective study/	931 667
44	Prospective study/	609 678
45	Randomized controlled trials/	180 820
46	44 not 45	603 226
47	Cohort analysis/	587 837

SUPPLEMENTAL TABLE 4 Continued

Number	Searches	Results
48	(Cohort adj (study or studies)).mp.	305 660
49	(Case control adj (study or studies)).tw.	133 568
50	(follow up adj (study or studies)).tw.	68 573
51	(observational adj (study or studies)).tw.	166 790
52	(epidemiologic\$ adj (study or studies)).tw.	109 791
53	(cross sectional adj (study or studies)).tw.	217 731
54	39 or 40 or 41 or 42 or 43 or 46 or 47 or 48 or 49 or 50 or 51 or 52 or 53	2 743 967
55	38 or 54	9 443 089
56	3 and 29 and 55	4101
57	animal/	1 957 779
58	human/	22 185 969
59	57 not (57 and 58)	1 486 477
60	56 not 59	4101
61	limit 60 to (books or chapter or conference abstract or conference paper or "conference review")	471
62	60 not 61	3630

Embase Classic + Embase, 1947 to 2020, week 25.

SUPPLEMENTAL TABLE 5 Full Electronic Search Strategy From Cochrane Central Register of Controlled Trials

Number	Searches	Results
1	Mucocutaneous Lymph Node Syndrome/	101
2	(kawasaki disease or kawasaki syndrome or mucocutaneous lymph node syndrome or viral mucocutaneous lymphadenopathy).tw,kw.	321
3	or/1-2	322
4	exp mortality/	12 988
5	(death rate? or mortalit*).tw,kw.	83 055
6	exp myocardial infarction/	10 702
7	(cardiovascular stroke? or heart attack? or myocardial infarct*).tw,kw.	31 655
8	exp Heart Failure/	9253
9	(cardiac failure? or heart decompensation or heart failure?).tw,kw.	29 727
10	exp Hypertension/	17 748
11	(high blood pressure? or hypertension).tw,kw.	57 375
12	exp coronary artery disease/	6300
13	(coronary arteriosclerosis# or coronary artery disease? or coronary atheroscleroses or coronary atherosclerosis).tw,kw.	15 029
14	exp Stroke/	9400
15	(stroke? or apoplexy or brain vascular accident? or cva or cvas or cerebrovascular accident? or cerebrovascular apoplexy).tw,kw.	58 053
16	Cerebral Revascularization/	59
17	exp myocardial revascularization/	9134
18	(revascularization? or "ec ic arterial bypass*" or extracranial intracranial arterial bypass* or "sta mca bypass*" or internal mammary artery implantation).tw,kw.	11 380
19	exp infection/	23 940
20	infection?.tw,kw.	101 155
21	exp eczema/	1034
22	eczema*.tw,kw.	3273
23	exp Asthma/	11 454
24	asthma*.tw,kw.	34 393
25	exp psoriasis/	3266
26	(palmoplantaris pustulosis or psoriasis# or "pustular psoriasis of palms and soles" or "pustulosis palmaris et plantaris" or "pustulosis of palms and soles").tw,kw.	7809
27	exp arthritis/	15 271
28	(inflammat* adj3 arthriti*).tw,kw.	1263
29	or/4-28	385 971
30	3 and 29	86

EBM Reviews, Cochrane Central Register of Controlled Trials, May 2020; EBM Reviews, Cochrane Database of Systematic Reviews, 2005 to June 17, 2020.

SUPPLEMENTAL TABLE 6 Examples of Potential Noncardiac Diseases From Literature Review and Expert Opinion

Rheumatoid arthritis
Juvenile idiopathic arthritis
Systemic lupus erythematosus
Psoriasis
Psoriatic arthritis
Inflammatory bowel diseases
Celiac disease
Hypothyroidism
Type 1 diabetes
Infections (bacterial infections)
Sensorineural hearing loss
Eczema
Allergies
Malignancy

SUPPLEMENTAL TABLE 7 All Mortality Studies in the Kawasaki Disease (KD) Population

Source	Study Design	Population	Relevant Selection Criteria	Median Age (IQR) at KD Diagnosis	Ethnicity, n (%)	Coronary Artery Aneurysms, n (%)	Median Duration F/U	Results	Increased Risk of Death in KD compared with non-KD patients	Bias (NOS: 0–9, JBI: 0–8)
Cumulative mortality per period										
Suzuki, 1988, Japan ⁸⁶	RC	Total: 36 KD patients; group 1: 23 KD patients with MI; group 2: 23 patients without MI but obstructive lesions on CA	KD patients with history of MI or CAA	Mean: group 1: 19.0 (23.6) mo; group 2: 25.9 (23.4) mo	NR	36 (100)	Mean 55.8 (24.4) mo	Group 1: 4 deaths (17.4%); group 2: no deaths	Unclear	NOS: 6
Tatara, 1989, Japan ⁸⁸	RC	30 KD patients	KD patients with coronary artery obstruction on CA	NR	NR	30 (100)	Mean 13.1 y	1 death (3.3%)	Unclear	NOS: 6
Akagi, 1992, Canada ⁸⁸	RC	583 KD patients	—	Mean 3.1 (2.7) y	Caucasian: 321 (55), Asian: 90 (15), Black: 50 (9), unknown: 122 (21)	Giant CAA: 80 (13.7), moderate-sized CAA: 44 (55.0), dilation: 14 (17.5)	Mean 4.0 (3.6) y	3 deaths (0.5%)	Unclear	NOS: 6
Kato, 1996, Japan ⁸⁹	RC	594 KD patients	KD patients with no treatment with IVIG	Mean 15 mo	NR	Overall: 146 (24.6); giant CAA: 26 (4.4)	Mean 13.6 y	5 deaths (0.8%) from 3 mo to 7 y after KD onset (mean 4.3 [1.6] y)	Unclear	NOS: 6
Miyagawa, 1998, Japan ⁹⁰	RC	80 KD patients	KD patients with CAA	Mean 2.3 (2.2) y	NR	90 (100)	8.8 (1.2) y	1 death (1.1%)	Unclear	NOS: 7
Lue, 2004, Taiwan ⁹²	CS	8339 KD patients	—	85% were <5 y of age	NR	2152 (25.8)	24 y	13 deaths (0.16%)	Unclear	JBI: 2
Tsuda, 2004, Japan ⁹³	CS	244 KD patients	KD patients who underwent CABG <20 y old for coronary stenosis	Age of onset: <5 y old: 197 (84%)	NR	NR	Follow-up interval from CABG to last visit: 5 y (7 d–25 y)	15 deaths (6.1%); 14 deaths occurred late (not in operative period) of which 9 were sudden.	Unclear	JBI: 3
Dadani, 2005, United States ⁸⁴	PC	Total: 18 KD patients; group 1: 9 patients with CAA present; group 2: 9 patients with no CAA	KD patients at least 1 y out from diagnosis	Mean: group 1: 3.2 (2.3) y; group 2: 5.2 (2.2) y	NR	9 (50)	Mean: group 1: 12.4 (4.9) y; group 2: 4.8 (3.0) y	1 sudden death (6.6%)	Unclear	NOS: 5
Chiang, 2013, Taiwan ⁴²	RC	8148 KD patients	—	Mean 2.16 (0.02) years	NR	620 (7.6)	7.7 y (IQR: 5.6–9.8)	20 deaths (0.25%). Mean time from KD diagnosis to death 2 (2.5) years	Unclear	NOS: 6
Holve, 2014, United States ⁴⁵	RC	KD: 546 patients; general population comparators: 2219 patients	KD patients at least 1 y out from diagnosis and age of ≥15 y old at time of study	NR	Caucasian: 202 (37), Hispanic: 98 (17.9), Asian: 101 (18.5), Black: 71 (13), multiracial/other: 18	None: 436 (79.9); transient ectasia: 19 (3.5); persistent ectasia: 3 (0.5); transient caa: 23	Mean 14.9 y	0 deaths	No	NOS: 8

SUPPLEMENTAL TABLE 7 Continued

Source	Study Design	Population	Relevant Selection Criteria	Median Age (IQR) at KD Diagnosis	Ethnicity, n (%)	Coronary Artery Aneurysms, n (%)	Median Duration F/U	Results	Increased Risk of Death in KD compared with non-KD patients	Bias (NOS: 0–9, JBI: 0–8)
Noto, 2014, Japan ⁸⁵	RC	58 KD patients	—	NR	(3.3), unknown: 54 (9.9)	(4.2); persistent caa: 25 (4.6); unknown: 40 (7.3)	Mean 14.7 (5.5) y	0 deaths	No	NOS: 6
Wei, 2015, China ⁸⁶	RC	38 KD patients	KD patients with persistent CAA during the study period	2.5 y	NR	38 (100)	4.1 y	7 deaths (18%); 3 were due to MI, 2 were sudden deaths, 2 were rupture of giant CAA	Unclear	NOS: 5
Jang, 2015, Korea ⁸⁶	RC	239 KD patients	KD patients with giant CAA (>6 mm)	3 y (range: 2 mo–16 y)	NR	239 (100)	7.7 y (range: 2 mo–21 y)	5 deaths (2%); median time from KD onset to death was 3 mo (range: 1 mo–3.3 y)	Unclear	NOS: 5
Friedman, 2016, United States ⁸⁴	RC	500 KD patients	KD patients with CAA at any time in their illness	<1 y old: 167 (33%); 1–4 y old: 235 (47%); ≥ 5 y old: 98 (20%)	Asian: 112 (22)	500 (100); giant CAA: 64 (15)	11.7 y	3 deaths (0.01%)	Unclear	NOS: 7
Gillebert, 2010, Belgium ⁸⁸	RC	35 KD patients	—	1.9 y	NR	Left coronary involvement: 94%; right coronary involvement: 58%; both right and left coronary involvement: 54%	4 y	0 deaths	No	NOS: 6
Wu, 2017, Taiwan ¹⁰¹	RC	6690 KD patients	—	16 mo	NR	920 (14)	Mean: 10.45 (3.02) y	10 (0.15%) deaths	Unclear	NOS: 6
Jeong, 2018, Korea ⁵¹	RC	20 KD patients	KD patients who underwent CABG	3 y (range: 0–7)	NR	20 (100)	Mean: 59.5 (48.5) mo	0 deaths	No	NOS: 5
Fukazawa, 2018, Japan ⁸⁷	RC	209 KD patients	KD patients with giant CAA (≥8 mm)	35 mo (range: 1–158 mo)	NR	209 (100)	Mean: 26 (13) mo	Total: 12 (5.7%) deaths, 10 deaths occurred within the first year of KD onset	Unclear	NOS: 6
Survival analyses										
Kitamura, 1994, Japan ⁸²	RC	Total: 168 KD patients; group 1: 54 patients operated with SVG; group 2: 114 patients operated with ITA	KD patients who underwent myocardial revascularization	NR. Mean age at time of operation: 10.6 (8.1) y	NR	161 (94.7)	NR. 50% of patients followed between 100–140 mo. Maximum observation of 12 y	With ITA: 98.7% (1.2) survival at 10 y; without ITA (SVG): 81.6% (7.0) survival at 10 y	Unclear: KD patients with ITA grafts had higher survival than KD patients with SVG	NOS: 6

SUPPLEMENTAL TABLE 7 Continued

Source	Study Design	Population	Relevant Selection Criteria	Median Age (QR) at KD Diagnosis	Ethnicity, n (%)	Coronary Artery Aneurysms, n (%)	Median Duration F/U	Results	Increased Risk of Death in KD compared with non-KD patients	Bias (NOS: 0–9, JBI: 0–8)
Yoshikawa, 2000, Japan ³³	RC	100 KD patients	for CAA, either with SVG or IFA KD patients with obstructive CAA	NR, Mean age at operation: 10 (5) y	NR	100 (100)	Mean: 6.7 (4.5) y	Total: 2 (0.2%) deaths. 98% survival at 3 y. 97.6% at 10 y.	Unclear	NOS: 5
Kitamura, 2009, Japan ³⁴	RC	114 KD patients	KD patients who underwent CABG for CAA	1 y	NR	114 (100)	19 y	Total: 5 (4%) deaths. 98% (95% CI: 95–100) survival at 10 y, 95% (88–98) at 20 y, 95% (88–98) at 25 y	Unclear	NOS: 7
Tsuda, 2011, Japan ³⁵	RC	60 KD patients	KD patients with an acute MI from 1976–2007	1.4 (0.19–12) y	NR	Total: 60 (100): 50 (85%) had giant CAA	Median from initial MI: 16 y	Total: 15 (25%) died at a median age of 16 y: 84.6% (95% CI, 72.8–91.8) survival at 10 y, 79.4% (65.9–88.4) at 20 y, and 62.7% (44.6–77.9) at 30 y; Cox Proportional HR: patients with LVEF < 45% had lower survival	Unclear: KD patients with low LVEF (<45%) had lower survival than those with higher LVEF	NOS: 6
Suda, 2011, Japan ⁴⁰	RC	76 KD patients	KD patients with giant CAA	Mean: 2.9 (2.9) y	NR	76 (100)	19 y (Range: 2 mo – 36 y)	Total: 8 (10.5%) deaths: 95% survival at 10 y, 88% at 20 y, and 88% at 30 y	Unclear	NOS: 6
Tsuda, 2014, Japan ³⁶	RC	245 KD patients	KD patients with giant CAA	24 mo	NR	245 (100)	20 y	Total: 15 (6%) deaths, median interval from KD to death: 11 y; overall: 97% (95% CI: 94–99) at 10 y, 95% (91–98) at 20 y, 90% (84–93) at 30 y. The survival rate for bilateral giant CAA was significantly lower than unilateral	Unclear: KD patients with bilateral giant CAA had lower survival than those with unilateral giant CAA	NOS: 6

SUPPLEMENTAL TABLE 7 Continued

Source	Study Design	Population	Relevant Selection Criteria	Median Age (IQR) at KD Diagnosis	Ethnicity, n (%)	Coronary Artery Aneurysms, n (%)	Median Duration F/U	Results	Increased Risk of Death in KD compared with non-KD patients	Bias (NOS: 0–9, JBI: 0–8)
Lin, 2015, Taiwan ⁴⁵	RC	1073 KD patients	—	1.59 y	NR	Total: 40.6% in acute febrile stage, of which 196 (18.3) had CAA that persisted beyond 1 mo	5.63 y	giant CAA at 30 y (87% vs 96%) Total: 4 (0.4%) deaths: 98% survival at 10 y, 98% survival at 15 y	Unclear	NOS: 5
Hoshino, 2015, Japan ⁵³	RC	20 KD patients	KD patients with systemic artery aneurysms from 1980–2013	6 mo (range: 1–20 mo)	NR	NR	18 y	Total: 4 (20%) deaths: 77% survival at 20 y	Unclear	NOS: 5
Chih, 2016, Taiwan ³⁸	RC	78 KD patients	KD patients with CAA ≥ 4 mm	1.59 y	NR	Giant CAA (≥ 8 mm): 27 (65); medium CAA (4–8 mm): 51 (65)	6.67 y	Total: 4 deaths: 14.8% mortality in patients with giant CAA, 5-y survival rate: 92%, 35-y survival rate: 69%	Unclear	NOS: 5
Tsuda, 2017, Japan ⁴⁷	RC	214 KD patients	KD patients with at least 1 CAA in a CA <100 d after KD onset	23 mo	NR	214 (100); small: 56; medium: 62; large: 96	16.8 y	30-y survival: large CAA: 94% (95%CI: 84–98); medium CAA: 96% (77–99); no significant difference between groups)	Unclear: No significant difference in survival between large and medium CAA groups	NOS: 5
Tadokoro, 2019, Japan ³⁹	RC	92 KD patients: single CABG: 53 patients; multiple CABG: 49 patients	KD patients who underwent CABG	NR, Mean patient age at surgery was 14.9 (10.4) y	NR	92 (100)	13 y (range: 1 mo–35 y)	6 (6.5%) deaths after CABG, interval range from 2 mo–11 y. Single-CABG Survival: 97.9% at 5 y, 92.8% at 10 y, 20 y, and 30 y. Multiple-CABG Survival: 95.1% at 5 y, 91.0% at 10 y, 20 y, and 30 y.	Unclear: No difference between single/multiple CABG	NOS: 7

Risk of bias was evaluated with the NOS for case-control or cohort studies, and was evaluated with the JBI checklist for cross-sectional studies. CA, coronary angiography; CC, case control; CI, confidence interval; CS, cross-sectional; F/U, follow-up; HR, hazard ratio; IQR, interquartile range; ITA, internal thoracic artery; IWG, intravenous immunoglobulin; LVEF, low ventricular ejection fraction; NR, not reported; PC, retrospective cohort; RC, retrospective cohort; SVG, saphenous vein graft. —, not applicable.

SUPPLEMENTAL TABLE 8 Nakamura Retrospective Cohort Studies: All-Cause Standardized Mortality Ratios (SMRs) Among KD Patients in Japan

Year of Publication (Ref)	End Date of Study	Mean Duration From KD (y)	SMR (95% CI)	Conclusion About Long-term Mortality
1996 ^a (25)	December 1992	5.7	Entire study period: all: 1.56 (0.94–2.43), males: 1.78 (0.97–2.99), females: 1.16 (0.38–2.71)	No difference compared with general population
1998 (26)	December 1994	7.6	Entire study period: all: 1.35 (0.82–2.08), males: 1.45 (0.79–2.44), females: 1.15 (0.42–2.52)	No difference compared with general population
2000 (27)	December 1997	10.7	Entire study period: all: 1.35 (0.87–2.00), males: 1.56 (0.94–2.44), females: 0.95 (0.35–2.07). After acute phase with cardiac sequelae: all: 2.01 (0.74–4.37), males: 2.77 (1.02–6.04), females: 0	The mortality rate among males with cardiac sequelae is statistically significantly increased compared with general population
2002 (28)	December 1999	12.6	Entire study period: all: 1.25 (0.84–1.85), males: 1.32 (0.82–2.10), females: 1.12 (0.52–2.30). After acute phase with cardiac sequelae: all: 1.72 (0.70–3.95), males: 2.35 (0.96–5.39), females: 0	The mortality rate among males with cardiac sequelae is higher but not statistically significant
2005 (29)	December 2001	14.6	Entire study period: all: 1.15 (0.77–1.66), males: 1.17 (0.71–1.80), females: 1.11 (0.51–2.11). After acute phase with cardiac sequelae: all: 1.45 (0.53–3.16), males: 1.95 (0.71–4.25), females: 0 (0–3.35)	The mortality rate among males with cardiac sequelae is higher but not statistically significant
2008 (30)	December 2004	17.6	Entire study period: all: 1.14 (0.80–1.57), males: 1.24 (0.79–1.80), females: 0.92 (0.42–1.75). After acute phase with cardiac sequelae: all: 1.92 (0.92–3.54), males: 2.55 (1.23–4.70), females: 0 (0–2.88)	The mortality rate among males with cardiac sequelae is statistically significantly increased compared with general population
2013 (31)	December 2009	22.6	Entire study period: all: 1.00 (0.73–1.34), males: 1.09 (0.76–1.52), females: 0.81 (0.40–1.45). After acute phase with cardiac sequelae: all: 1.86 (1.02–3.13), males: 2.27 (1.21–3.87), females: 0.56 (0.01–3.14)	The mortality rate among all patients with cardiac sequelae and, in particular, males with cardiac sequelae is statistically significantly increased compared with general population

CI, confidence interval; SMR, standardized mortality ratio

^a This study included 6585 patients, whereas the remaining studies included 6575 patients. Reasons for the exclusion of the 10 patients for subsequent publications and analyses were not described.

SUPPLEMENTAL TABLE 9 Mortality in KD Patients With Known Coronary Artery Aneurysms (CAAs)

Source	Population	Median Age (IQR) at Diagnosis	Median Duration F/U	Results
Suzuki, 1988, Japan ⁸⁶	Total: 36 KD patients with history of MI/CAA; group 1: 23 KD patients with MI; group 2: 23 patients without MI but obstructive lesions on CA	Mean: group 1: 19.0 (23.6) mo; group 2: 25.9 (23.4) mo	Mean 55.6 (24.4) mo	Group 1: 4 deaths (17.4%); group 2: No deaths
Tatara, 1989, Japan ⁸⁹	30 KD patients with coronary artery obstruction	NR	Mean 13.1 y	1 death (3.3%)
Miyagawa, 1998, Japan ⁹⁰ Tsuda, 2004, Japan ⁹³	90 KD patients with CAA 244 KD patients with CABG <20 y of age	Mean 2.3 (2.2) y Age of onset: <5 y old: 197 (84%)	8.8 (1.2) y Follow-up interval from CABG to last visit: 5 y (7 d–25 y)	1 death (1.1%) 15 deaths (6.1%): 14 dates occurred late (not in operative period) of which 9 were sudden.
Wei, 2015, China ⁹⁸	38 KD patients with persistent CAA	2.5 y	4.1 y	7 deaths (18%): 3 were due to MI, 2 were sudden deaths, 2 were rupture of giant CAA
Jang, 2015, Korea ⁹⁶	239 KD patients with CAA>6 mm	3 y (range: 2 mo–16 y)	7.7 y (range: 2 mo–21 y)	5 deaths (2%) Median time from KD onset to death was 3 mo (range: 1 mo–3.3 y)
Friedman, 2016, United States ⁵⁴	500 KD patients with CAA	<1 y old: 167 (33%); 1–4 y old: 235 (47%); ≥ 5 y old: 98 (20%)	11.7 y	3 deaths (0.6%)
Gillebert, 2010, Belgium ⁹⁹ Jeong, 2018, Korea ⁵¹ Fukazawa, 2018, Japan ⁴⁷	35 KD patients 20 KD patients with CABG 209 KD patients with giant CAA (≥8 mm)	1.9 y 3 y (range: 0–7) 35 mo (range: 1–158 mo)	4 y Mean: 59.5 (48.5) mo Mean: 26 (13) mo	0 deaths 0 deaths Total: 12 (5.7%) deaths, 10 deaths occurred within the first year of KD onset
Kitamura, 1994, Japan ³²	Total: 168 KD patients with coronary procedures; group 1: 54 patients operated with SVG; group 2: 114 patients operated with ITA	NR. Mean age at time of operation: 10.6 (8.1) y	NR. 50% of patients followed between 100–140 mo. Maximum observation of 12 y	With ITA: 98.7% (1.2) survival at 10 y; without ITA (SVG): 81.6% (7.0) survival at 10 y
;Yoshikawa, 2000, Japan ³⁵	100 KD patients with obstructive CAA	NR. Mean age at operation: 10 (5) y	Mean: 6.7 (4.5) y	Total: 2 (2%) deaths: 99% survival at 3 y, 97.6% at 10 y.
Kitamura, 2009, Japan ³⁴	114 KD patients with CABG	1 y	19 y	Total: 5 (4%) deaths; 98% (95%CI, 93–100) survival at 10 y, 95% (88–98) at 20 y, 95% (88–98) at 25 y
Tsuda, 2011, Japan ³⁵	60 KD patients with acute MI from 1976–2007	1.4 (0.19–12) y	Median from initial MI: 16 y	Total: 15 (25%) died at a median age of 16 y; 84.6% (95%CI, 72.8–91.8) survival at 10 y, 79.4% (65.9–88.4) at 20 y, and 62.7% (44.6–77.9) at 30 y. Cox Proportional HR: patients with LVEF<45% had lower survival
Suda, 2011, Japan ⁴⁰	76 KD patients with giant CAA	Mean: 2.9 (2.9) y	19 y (range: 2 mo–36 y)	Total: 8 (10.5%) deaths: 95% survival at 10 y, 88% at 20 y, and 88% at 30 y
Tsuda, 2014, Japan ³⁶	245 KD patients with giant CAA	24 mo	20 y	Total: 15 (6%) deaths, median interval from KD to death: 11 y; overall:

SUPPLEMENTAL TABLE 9 Continued

Source	Population	Median Age (IQR) at Diagnosis	Median Duration F/U	Results
Chih 2016, Taiwan ³⁸	78 KD patients with CAA \geq 4 mm	1.59 y	6.67 y	97% (95%CI, 94–99) at 10 y, 95% (91–98) at 20 y, 90% (84–93) at 30 y. The survival rate for bilateral giant CAA was significantly lower than unilateral giant CAA at 30 y (87% vs 96%). Total: 4 deaths (5.1%): 14.8% mortality in patients with giant CAA. 5-y survival rate: 92%; 35-y survival rate: 69%
Tsuda, 2017, Japan ³⁷	214 KD patients with at least 1 CAA in <100 d from KD onset	23 mo	16.8 y	Total: 5 deaths (2.3%): 30-y survival: large CAA: 94% (95%CI, 84–98); medium CAA: 96% (77–99); no significant difference between groups
Tadokoro, 2019, Japan ³⁹	92 KD patients with CABG; single CABG: 53 patients; multiple CABG: 49 patients	NR. Mean patient age at surgery was 14.9 (10.4) y	13 y (range: 1 mo–35 y)	6 (6.5%) deaths after CABG, interval range from 2 mo–11 y; single-CABG Survival: 97.9% at 5 y, 92.8% at 10 y, 20 y, and 30 y; multiple-CABG Survival: 95.1% at 5 y, 91.0% at 10 y, 20 y, and 30 y.

CA, coronary angiography; CI, confidence interval; F/U, follow-up; IQR, interquartile range; ITA, internal thoracic artery; SVG, saphenous vein graft; NR, not reported.

SUPPLEMENTAL TABLE 10 Survival in KD Patients With Known Coronary Artery Aneurysms (CAAs) Over Follow-Up Time

Years of Follow-Up	Study	Survival Percentage (95%CI)
5 y	Yoshikawa, 2000 ³³	99% at 3 y
	Chih, 2016 ³⁸	92%
10 y	Tadokoro, 2019 ³⁹	97.9% in KD patients with single-CABG; 95.1% in KD patients with multiple-CABG
	Kitamura, 1994 ³²	98.7% in KD patients with ITA, 81.6% in KD patients without ITA
	Yoshikawa, 2000 ³³	97.6%
	Kitamura, 2009 ³⁴	98% (93%–100%)
	Tsuda, 2011 ³⁵	84.6% (72.8%–91.8%)
	Suda, 2011 ⁴⁰	95%
	Tsuda, 2014 ³⁶	97% (94%–99%)
20 y	Tadokoro, 2019 ³⁹	92.8% in KD patients with single-CABG; 93.0% in KD patients with multiple-CABG
	Kitamura, 2009 ³⁴	95% (88%–98%)
	Tsuda, 2011 ³⁵	79.4% (65.9%–88.4%)
	Suda, 2011 ⁴⁰	88%
	Tsuda, 2014 ³⁶	95% (91%–98%)
25 y	Tadokoro, 2019 ³⁹	92.8% in KD patients with single-CABG; 91.0% in KD patients with multiple-CABG
	Kitamura, 2009 ³⁴	95% (88%–98%)
	Tsuda, 2011 ³⁵	62.7% (44.6%–77.9%) ^a
30 y	Suda, 2011 ⁴⁰	88%
	Tsuda, 2014 ³⁶	90% (84%–93%)
	Tsuda, 2017 ³⁷	94% (84%–98%) in KD patients with large CAA; 96% (77%–99%) in KD patients with medium CAA
	Tadokoro, 2019 ³⁹	92.8% in KD patients with Single-CABG; 91.0% in KD patients with multiple-CABG
35 y	Chih, 2016 ³⁸	69%

CI, confidence interval; ITA, internal thoracic artery.

^a Patients with CAA and acute MI.

SUPPLEMENTAL TABLE 11 All Studies Evaluating Cardiovascular Outcomes in the KD Population

Source	Study Design	Population	Selection Criteria	Median Age at KD Diagnosis	Ethnicity, n (%)	CAA, n (%)	Median Duration F/U (y)	Cardiac Outcome(s)	Results	Increased Risk in KD compared with Comparators (Yes, No, Unclear)	Bias (NOS: 0-9, JBI: 0-8)
Primary analyses: descriptive or univariable analyses											
Kato, 1982, Japan ¹⁰⁵	RC	43 KD patients	Diagnosed with CAA in the acute phase of illness	Mean 19 mo	NR	43 (100)	Mean 4.3 y	MI, exercise testing	MI: 6 patients (14%) had MI, 1 patient died and 5 recovered. Exercise testing: 5 (58%) of 13 patients with ongoing abnormal CA showed abnormal exercise testing. 0 (0%) of 6 patients with regressed CAA had normal exercise testing	Unclear	NOS: 5
Novelli, 1984, UK ⁵⁶	RC	18 KD patients	—	Mean 15.3 mo (range: 4 mo–3 y 3 mo)	White: 14 (78), Black: 2 (11), Asian: 2 (11)	6 (33)	Mean 12.3 mo	HTN, MI	HTN: 8 (44%), 6 patients were mild without treatment; MI: 1 (6%)	Unclear	NOS: 5
Suzuki, 1988, Japan ⁸⁸	RC	Total: 36 KD patients; group 1: 23 KD patients with MI; group 2: 23 patients without MI but obstructive lesions on CA	KD patients with history of MI or CAA	Mean: group 1: 19.0 (23.6) mo; group 2: 25.9 (23.4) mo	NR	36 (100)	Mean 55.6 (24.4) mo	HF, angina, cardiac interventions	HF: 1 patient in group 1, 0 patient in group 2. Angina: 2 patients in group 1, 0 patient in group 2. Cardiac interventions: 3 patients in group 1, 5 patients in group 2	Unclear. Children with clinical MIs had worse cardiac outcomes than those without MIs	NOS: 6
Tatara, 1989, Japan ⁸⁹	RC	30 KD patients	KD patients with coronary artery obstruction on CA	NR	NR	30 (100)	Mean 13.1 y	MI	MI: 8 patients (27%) had clinical symptoms of MI; 7 patients had positive stress testing	Unclear	NOS: 6
Newburger, 1991, Japan ⁵⁷	RC	105 KD patients; 46 KD patients were compared with parents.	—	9 (8.6%) were <2 y old	White: 95 (90), Black: 6 (6), Asian: 4 (4)	NR	NR. 30 patients had lipid profiles at >3 y	HLD - Lipid values were adjusted for age and sex	HLD: Total cholesterol at time period 1 (<10 d from KD) was significantly lower than all periods. HDL was depressed <10 d from KD and then increased significantly with time, with the highest at period 6 (>3 y from	Yes. HDL values were significantly lower than expected according to age and sex.	NOS: 5

SUPPLEMENTAL TABLE 11 Continued

Source	Study Design	Population	Selection Criteria	Median Age at KD Diagnosis	Ethnicity, n (%)	CAA, n (%)	Median Duration F/U (y)	Cardiac Outcome(s)	Results	Increased Risk in KD compared with Comparators (Yes, No, Unclear)	Bias (NOS: 0-9; JBI: 0-8)
Akagi, 1992, Canada ⁸⁸	RC	583 KD patients	—	Mean 3.1 (2.7) y	White: 321 (55), Asian: 90 (15), Black: 50 (9), unknown: 122 (21)	Giant aneurysm: 80 (13.7); moderate-sized aneurysm: 44 (55.0); dilation: 14 (17.5)	Mean 4.0 (3.6) years	MI, CABG	KD, but when adjusted, KD patients had HDL values that were significantly lower than normal. Subgroup compared with parents: adjusted total cholesterol not significantly different from parent(s), but had significantly lower HDL than parents MI: 9 patients (1% of total cohort, 11% with CAA, 100% with giant CAA). Mean interval from KD was 1.5 (2.1) years. CABG: 2 patients	Unclear. Descriptively, KD patients with giant CAA had more MIs compared with those who did not have giant CAA	NOS: 6
Kitamura, 1994, Japan ³²	RC	Total, 168 KD patients; group 1: 54 patients operated with SVG; group 2: 114 patients operated with ITA	KD patients who underwent myocardial revascularization for CAA, either with SVG or ITA	NR. Mean age at time of operation: 10.6 (8.1) y	NR	161 (94.7)	NR. 50% of patients followed between 100–140 mo. Maximum observation of 12 y	Post-operative MI, arrhythmia, angina	MI: Mild ischemia in 14% by stress ECHO. Clinical MI occurred in 2%. Arrhythmia: ventricular arrhythmia in 1%. Angina: 7% of patients. No difference in nonfatal cardiac events between ITA/SVG groups	No. No difference in nonfatal cardiac events between 2 KD groups (ITA/SVG)	NOS: 6
Kato, 1996, Japan ⁸⁹	RC	594 KD patients	KD patients with no treatment with IVIG	Mean 15 mo	NR	Overall: 146 (24.6); giant CAA: 26 (4.4)	Mean 13.6 y	Systemic AA, MI, CABG	Systemic AA: 13 (2%). MI: 11 (2%) of which 8 patients had giant CAA. 4 patients had MI within 1 y from diagnosis. CABG: 7 (1%)	Unclear	NOS: 6
Noto, 2001, Japan ⁵⁹	CS	20 KD patients and 20 healthy controls (age and sex matched)	KD patients with CAA	NR	NR	20 (100)	Mean 9.8 (4) y	HLD, HTN, cIMT	HTN: No significant difference in systolic or diastolic BPs between groups. HLD: No significant difference in TG, total cholesterol,	HLD, HTN: No IMT: Yes	JBI: 7

SUPPLEMENTAL TABLE 11 Continued

Source	Study Design	Population	Selection Criteria	Median Age at KD Diagnosis	Ethnicity, n (%)	CAA, n (%)	Median Duration F/U (y)	Cardiac Outcome(s)	Results	Increased Risk in KD compared with Comparators (Yes, No, Unclear)	Bias (NOS: 0-9, JBI: 0-8)
Murakami, 2003, ⁸⁵ Japan ⁸⁵	CS	60 KD patients	—	NR	NR	20 (33)	Minimum 15 y from diagnosis	MI, angina, CABG	HDL between groups. TG, total cholesterol, LDL, HDL all within normal limits in KD. IMT: Significantly higher in KD than in controls (4.11 [0.86] mm vs 2.94 [0.91] mm)	Unclear	JBI: 4
Tsuda, 2004, ⁸⁶ Japan ⁸⁵	CS	244 KD patients	KD patients who underwent CABG <20 y old (84%)	Age of onset: <5 y old: 187 (84%)	NR	NR	Follow-up interval from CABG to last visit: 5 y (7 d–25 y)	Re-CABG, MI, HF	MI: 4 (7%); angina: 9 (15%); CABG/PCI: 3 (5%) Re-CABG: 14 patients. MI: 70 patients (28%) had a MI prior to CABG. HF: 205 patients classified at NYHA I, 8 patients classified at NYHA II. 138 patients had ≥50% LVEF, 13 had 40–50%, and 8 patients had <40%	Unclear	JBI: 3
Dadlani, 2005, ⁸⁷ United States ⁸⁴	PC	Total: 18 KD patients; group 1: 9 patients with CAA present; group 2: 9 patients with no CAA	KD patients at least 1 y out from diagnosis	Mean: group 1: 3.2 (2.3) y; group 2: 5.2 (2.2) y	NR	9 (50)	Mean: group 1: 12.4 (4.9) y; group 2: 4.8 (3.0) y	Coronary artery events (defined as MI or sudden death)	Coronary artery events: no events in group 1 or group 2	No	NOS: 5
Levy, 2005, ⁸⁸ Canada ¹⁰⁷	RC	Total: 22 KD patients; group 1: 13 KD patients on warfarin; group 2: 9 KD patients not on warfarin	KD patients with giant CAA	Group 1: 3.7 y (range: 0.3–17.1); group 2: 3.5 y (range: 1.1–10.5)	NR	22 (100)	Group 1: 6 y (range 1–17.5); group 2: 14 y (range 5.3–20.5)	MI, late ischemia (monitored by exercise testing and myocardial perfusion imaging)	MI: 3 patients (14%) had early MIs at 1, 3, and 8 mo after diagnosis. Late ischemia: 1 patient in Group 2 had reversible perfusion defects (mean follow-up 10.5 y)	No. There was no difference in ischemic events between KD patients on and off warfarin	NOS: 6
Tsuda, 2006, ⁸⁹ Japan ¹⁰⁸	CS	30 KD patients	KD patients with CAA and obstetric outcomes	NR	NR	30 (100)	Median maternal age at delivery: 27 y (range 18–35 y)	CABG, MI, HF	CABG: 4 patients prior to pregnancy. MI: 0 (0%) patients. HF: all patients were classified as NYHA Class I	Unclear	JBI: 3
	CS	35 KD patients; group A: 9	KD patients who never smoked	Mean 2.7 (0.3) y	NR	9 (26%) with CAA, 6 (17%)	Mean 24.1 y	HLD, endothelial dysfunction	HLD: no difference in TG, HDL, LDL, TC/HDL ratio	HTM: No; endothelial dysfunction: Yes	JBI: 4

SUPPLEMENTAL TABLE 11 Continued

Source	Study Design	Population	Selection Criteria	Median Age at KD Diagnosis	Ethnicity, n (%)	CAA, n (%)	Median Duration FU (y)	Cardiac Outcome(s)	Results	Increased Risk in KD compared with Comparators (Yes, No, Unclear)	Bias (NOS: 0-9, JBI: 0-8)
Nihoshi, 2007, ⁶² Japan ⁶²		patients with CAA; group B: 6 patients with transient CAA; group C: 20 patients without CAA; 36 Non-KD controls				with transient CAA		(FMD), arterial stiffness (baPWV)	between KD groups and controls. Endothelial dysfunction: %FMD and FMD/ED ratio was markedly reduced in KD group compared with controls. FMD was reduced more in group A compared with group C. Arterial stiffness: baPWV values were significantly increased in male KD groups compared with male controls		
Ou, 2009, ⁶³ Taiwan ⁶³	CS	Total: 119 KD patients; group 1: 55 patients with persistent CAA; group 2: 64 patients without CAA	KD patients at least 1 y from diagnosis	Mean: group 1: 1.85 y (range 0.51–5.74); group 2: 2.09 y (range 0.63–6.24)	NR	55 (46)	Mean: group 1: 2.46 y (range 0.71–4.62); group 2: 2.52 y (range 1.12–4.92)	HLD, HTN	HTN: groups did not differ significantly between sBP and dBP. HLD: no difference in LDL-C, TG, total cholesterol. HDL levels were significantly different: group 1: 42.5 (SD: 10.23) mg/dL, group 2: 44.23 (10.91) mg/dL	HTN: No. HLD: unclear. KD patients with CAA have lower HDL levels	JBI: 6
Samada, 2010, ⁶⁸ Japan ⁶⁸	RC	48 KD patients	≥20 y old by 2005 and giant CAA	2.9 y (range: 0.3–12.8)	NR	48 (100)	NR. Median age at study was 25.2 (20–33.1) y	CABG, angina, MI	CABG: 9 patients, median 10.6 y (2.7–21.3) after diagnosis. Angina: 6 patients. MI: 14 patients (29%), all patients had either complete occlusion or stenosis	Unclear	NOS: 5
Chiang, 2013, ⁴² Taiwan ⁴²	RC	8148 KD patients	—	Mean 2.16 (0.02) y	NR	620 (7.6)	7.7 y (QR: 5.6–9.8)	MI, coronary artery complications (MI, angina, CAA)	MI: 0.2% (12 patients). Coronary artery complications: overall incidence was 11.53 per 1000 person-years. Mean time to coronary artery complications was 2.36 (11.6) months	Unclear	NOS: 6

SUPPLEMENTAL TABLE 11 Continued

Source	Study Design	Population	Selection Criteria	Median Age at KD Diagnosis	Ethnicity, n (%)	CAA, n (%)	Median Duration FU (y)	Cardiac Outcome(s)	Results	Increased Risk in KD compared with Comparators (Yes, No, Unclear)	Bias (NOS: 0-9, JBI: 0-8)
Cho, 2013, Korea ⁶⁴	CS	Group A: 19 KD patients with persistent/regressed CAA; group B: 49 KD patients without CAA; group C: 30 age- and sex-matched non-KD patients	—	NR	NR	19 (28) 8 had persistent CAA and 11 had regressed CAA	Mean: group A: 68.95 (31) mo; group B: 52.29 (27) mo	HTN, HLD, arterial stiffness (PWV)	HTN: no difference among the 3 groups. HLD: levels of TC, LDL, ppo-B were higher in group A than C. No difference between group A and B. No difference between ppo-A, HDL-C, and TG between groups. Arterial stiffness: PWV of KD patients were significantly higher in KD groups than group C	HTN: No. HLD: Unclear. Arterial stiffness: Yes	JBI: 4
Ishikawa, 2013, Japan ⁶⁰	CS	Group 1: 9 KD patients with CAA; group 2: 15 KD patients without CAA; group 3: 22 age- and sex-matched non-KD patients	1-5 y from KD diagnosis	Group 1: 3.3 y (range: 2.2-5.7); group 2: 3.3 y (1.9-4.3)	NR	9 patients	Group 1: 4.0 y (range 3.2-4.1); group 2: 2.0 y (1.0-3.4)	HTN, HLD, endothelial dysfunction (FMD)	HTN: No significant difference; HLD: No significant difference in TC, HDL, LDL, TC/HDL, TG; Endothelial dysfunction: %FMD was significantly lower in group 1 compared to group 2 and group 3. %FMD in group 2 was significantly lower than group 3	HTN, HLD, Endothelial dysfunction: Yes	JBI: 6
Wei, 2015, China ⁶⁶	RC	38 KD patients	KD patients with persistent CAA	2.5 y	NR	38 (100)	4.1 y	MI	MI: 6 patients (16%)	Unclear	NOS: 5
Gravel, 2015, Canada ⁴⁶	CS	Total: 250 KD patients; group 1: 133 KD patients with CAA; group 2: 117 KD patients without CAA	KD patients >6 y old and ≥ 1 y from diagnosis	NR	Group 1: Asian 65%, White 27%, other 8%. Group 2: Asian 47%, White 40%, other 13%	133 (53)	Mean: group 1: 7.5 (5.6) y; group 2: 6.8 (3.1) y	Exercise testing	Exercise testing: no difference in endurance time, maximum HR, BP responses. Proportion of abnormal perfusion scans was similar	No. There is no difference in exercise testing response according to CAA status	JBI: 8
Jang, 2015, Korea ⁶⁶	RC	239 KD patients	KD patients with giant CAA (>6 mm)	3 y (range: 2 mo-16 y)	NR	239 (100)	7.7 y (range: 2 mo-21 y)	MI, CABG, cardiac intervention	MI: 13 (5%) with a median age of onset of 1.9 y (range: 3 mo-5 y). CABG: 14 patients with a median age of 8.1 y (6 mo-22 y). Cardiac	Unclear	NOS: 5

SUPPLEMENTAL TABLE 11 Continued

Source	Study Design	Population	Selection Criteria	Median Age at KD Diagnosis	Ethnicity, n (%)	CAA, n (%)	Median Duration F/U (y)	Cardiac Outcome(s)	Results	Increased Risk in KD compared with Comparators (Yes, No, Unclear)	Bias (NOS: 0-9, JBI: 0-8)
Friedman, 2016, ¹⁶ United States ⁵⁴	RC	500 KD patients	KD patients with CAA	<1 y old: 167 (33%); 1-4 y old: 235 (47%); ≥5 y old: 98 (20%)	Asian: 112 (22)	500 (100); giant aneurysms: 64 (15)	11.7 y	MACE (MI, CABG, coronary interventions, cardiac death)	interventions: 22 patients at a median age of 7.7 y (3.9-13 y) MACE: Occurred in 24 patients at a median age of 3.5 y and median time of diagnosis of 1.5 y. Includes 4 symptomatic MI, 8 asymptomatic MI, 2 PCI, 6 CABG, 1 heart transplant, 3 cardiac deaths. Associated with higher CAA z-score at diagnosis (OR: 1.1) and lack of IVIG treatment (OR: 9.0)	Unclear	NOS: 7
Gilbert, 2017, ⁵⁵ Belgium ⁵⁶	RC	35 KD patients	—	1.9 y	NR	Left CAA: 94%; right CAA: 58%; both right and left CAA: 54%	4 y	MI	MI: 1 patient (3%)	Unclear	NOS: 6
Panihar, 2017, ⁵⁷ India ⁵⁸	CC	20 KD patients; age- and sex-matched controls	KD patients with transient CAA with at least 1 y from CAA resolution	NR	NR	20 had transient CAA	Mean: 53.7 mo	Arterial stiffness (cIMT, stiffness index)	Arterial stiffness: cIMT was similar in cases and controls and there was no significant difference in carotid artery stiffness index	No	NOS: 8
Gopalan, 2018, ⁵⁹ India ⁶⁰	PC	27 KD patients; age- and sex-matched comparators or closest sibling	—	NR	NR	10 (37) dilatations, 1 had CAA	Mean: 85.77 (14.12) mo	HLD, arterial stiffness (cIMT)	HLD: abnormal lipid profile was seen in 7 patients. There was no statistically significant difference in cholesterol, HDL, LDL, TG levels between KD patients with and without CAA at diagnosis. Arterial stiffness: mean cIMT was higher in KD patients than controls	HLD: Unclear; Arterial stiffness: Yes	NOS: 7
	RC	17 809 KD patients	—	NR	NR	1455 (8)	8 y (IQR: 4-11)			Unclear	NOS: 6

SUPPLEMENTAL TABLE 11 Continued

Source	Study Design	Population	Selection Criteria	Median Age at KD Diagnosis	Ethnicity, n (%)	CAA, n (%)	Median Duration F/U (y)	Cardiac Outcome(s)	Results	Increased Risk in KD compared with Comparators (Yes, No, Unclear)	Bias (NOS: 0-9, JBI: 0-8)
Coon, 2018, United States ⁴⁴					White: 7178 (40), Black: 3576 (20), Hispanic: 2529 (14), Asian: 1484 (8), other: 2359 (13), unknown: 683 (4)			Cardiac events (MI, cardiac arrest, heart failure, arrhythmia, CABG, cardiac interventions)	Cardiac events: 342 children experienced a cardiac event. 88% did so at the first year of diagnosis. Ischemic heart disease and/or heart failure accounted for 62% of events. Incidence rate: 19 per 1000 KD patients		
Primary analysis: linear/logistic regression											
Miyagawa, 1998, Japan ⁸⁰	RC	90 KD patients	KD patients with CAA	Mean 2.3 (2.2) y	NR	90 (100)	8.8 (1.2) y	Cardiac events (cardiac death, MI, CABG, unstable angina)	Total: 15 cardiac events (event rate: 17%: 1 death, 5 MI, 2 CABG, 7 unstable angina). Presence of thallium redistribution on SPECT was a multivariable independent predictor of a late cardiac event	Unclear	NOS: 7
Cheung, 2004, Hong Kong ⁵⁸	CS	Group 1: 37 KD patients with CAA; group 2: 29 KD patients without CAA; group 3: 36 healthy aged-matched children	KD patients \geq 1 y from diagnosis	NR	NR	37 (28.4%) with CAA, 21 had persistent CAA and 16 had CAA which regressed	Mean: group 1: 7.8 (3.7) y; group 2: 5.2 (2.4) y	HLD, HTN, arterial stiffness (PWV)	HLD: HDL was significantly lower in group 1 compared to group 2 or group 3. ApoA-I levels were significantly lower in group 1 compared to group 2 or group 3. There were no significant differences in TC or TG. HTN: No difference between the 3 groups. Arterial stiffness: PWV of group 1 was significantly higher than group 3. PWV of group 2 was significantly higher than group 3. There	HLD: Yes. HTN: No. Arterial stiffness: Yes	JBI: 6

SUPPLEMENTAL TABLE 11 Continued

Source	Study Design	Population	Selection Criteria	Median Age at KD Diagnosis	Ethnicity, n (%)	CAA, n (%)	Median Duration F/U (y)	Cardiac Outcome(s)	Results	Increased Risk in KD compared with Comparators (Yes, No, Unclear)	Bias (NOS: 0-9, JBI: 0-8)
Cheung, 2006, Hong Kong ⁶⁵	CS	Group 1: 26 KD patients with CAA; group 2: 24 KD patients without CAA; group 3: 22 healthy age-matched children	—	Mean: group 1: 1.5 (1.7) y; group 2: 2.7 (2.6) y	NR	26 patients	Mean: group 1: 7.4 (3.5) y; group 2: 5.8 (2.1) y	Arterial stiffness (cIMT, stiffness index, PWV), HLD, HTN	<p>was no significant difference between groups 1 and 2</p> <p>Arterial stiffness: stiffness index and brachioradial PWV differed significantly between groups, with group 1 having a higher stiffness index than group 2 and group 2 having a higher stiffness index than group 3. Group 1 had faster PWV than group 3. cIMT differed significantly between 3 groups with group 1 and 2 having greater cIMT than group 3. cIMT correlated with LDL, stiffness index, and PWV. HTN: No difference between groups. HLD: No difference in TC or HDL between groups. Group 1 had higher LDL compared with groups 2 and 3</p>	<p>Arterial Stiffness: Yes: HTN: No. HLD: Unclear</p>	JBI: 4
McCrindle, 2007, Canada ⁶¹	CC	52 KD patients and 60 healthy controls	10–20 y old KD patients	Mean: 4 (3) y	NR	Aneurysms: 37%; ectasia: 16%	Mean: 11.2 (0.37) y	Arterial stiffness (FMD, BAR), HTN, HLD,	<p>HTN: KD patients had lower SBP. No significant difference in 24-h ambulatory BP monitoring. HLD: KD patients had lower levels of ApoA-I. There was no significant difference in the remaining lipid profiles. Arterial stiffness: no significant difference in BAR. No</p>	<p>HTN, HLD, arterial Stiffness: No</p>	NOS: 5

SUPPLEMENTAL TABLE 11 Continued

Source	Study Design	Population	Selection Criteria	Median Age at KD Diagnosis	Ethnicity, n (%)	CAA, n (%)	Median Duration F/U (y)	Cardiac Outcome(s)	Results	Increased Risk in KD compared with Comparators (Yes, No, Unclear)	Bias (NOS: 0-9, JBI: 0-8)
Chen, 2017,		Australia ¹¹¹	CC	Australia cohort: 57 KD patients and 59 age and sex-matched controls. Singapore cohort: 78 KD patients and 78 age- and sex-matched controls	6–18 y old KD patients, at least 2 y from KD diagnosis	Mean: Australia: 3.5 (3.2) y; Singapore: 2.9 (2.4) y	Australia: White 73.7%, Asian 17.5%, other 1.8%, mixed: 7%. Singapore: Asian 100%	Australia: 32 (56.1); Singapore: 12 (15.4)	relationship noted with presence/absence of CAA Mean: Australia: 11.8 (5.9) y; Singapore: 6.9 (4.0) y	Retinal microvascular (marker of endothelial dysfunction)	Retinal NOS: 6
Primary analysis: survival analysis									microvasculature: KD patients from Singapore had larger retinal venules than controls. KD patients with CAA from Australia had larger retinal venules than controls but not significant. Retinal venular caliber increases were not associated with age, sex, time since KD, mean BP, or BMI	Unclear	NOS: 5
Yoshikawa, 2000, Japan ³³	RC	100 KD patients	KD patients with obstructive CAA	NR, Mean age at operation: 10 (5) y	NR	100 (100)	Mean 6.7 (4.5) y	Cardiac events (postoperative MI, repeat CABG, catheter intervention)	Cardiac events: event free rate was 70.5% at 10 y. Postoperative MI occurred in 15 patients, repeat CABG in 4 patients, catheter interventions in 7.	Unclear	NOS: 7

SUPPLEMENTAL TABLE 11 Continued

Source	Study Design	Population	Selection Criteria	Median Age at KD Diagnosis	Ethnicity, n (%)	CAA, n (%)	Median Duration F/U (y)	Cardiac Outcomes(s)	Results	Increased Risk in KD compared with Comparators (Yes, No, Unclear)	Bias (NOS: 0-9, JBI: 0-8)
Yamauchi, 2004, ⁴⁸ Japan ⁴⁸	Total: 21 KD patients; group S (short duration from KD diagnosis to CABG): 12 KD patients; group L (long duration from KD diagnosis to CABG): 9 KD patients	KD patients with CABG using arterial grafts	Mean 2.7 y; group S: 3.7 y (range: 1-7.7) from KD to CABG; group L: 13.9 y (range: 10-18) from KD to CABG	NR	85 (100)	Mean 5.8 (5.2) y	Cardiac events (heart failure, MI, valvular dysfunction)	Cardiac events: event free rate for group S was 100% and for group L was 66.7% during the follow-up period	Yes: KD patients who had a CABG closer to KD diagnosis had better prognosis	NOS: 5	
Suda, 2009, ⁴⁶ Japan ⁴⁶	85 KD patients	KD patients with giant CAA and on combination therapy of warfarin and aspirin for >3 mo	Mean 3.5 (3) y	NR	85 (100)	Mean 5.8 (5.2) y	Cardiac events (MI)	Cardiac events: event free rate was 92.7% at 6 mo, 92.7% at 1 y, 91.2% at 5 y, and 91.2% at 10 y. There was a total of 14 events and 10 (88%) occurred within the first year after KD onset	Unsure	NOS: 5	
Mueller, 2009,	Switzerland ⁴⁵	RC	Yes: Those with CAA >5 mm had more cardiac interventions	38 KD patients	KD patients with CAA	1.2 y (range 0.1-12.8)	NR	38 (100), 23 patients had ≤5 mm, 15 patients had >5 mm	8.5 y (range 0.5-24.8)	Cardiac interventions	Cardiac
	interventions: 4 patients with CAA >5 mm had stenosis/ ischemia and 2 required cardiac interventions. No patients with CAA <5 mm required cardiac interventions.										
	Event-free rate when stratified according to CAA (≤5 mm or >5 mm), >5 mm group had significantly										

SUPPLEMENTAL TABLE 11 Continued

Source	Study Design	Population	Selection Criteria	Median Age at KD Diagnosis	Ethnicity, n (%)	CAA, n (%)	Median Duration F/U (y)	Cardiac Outcome(s)	Results	Increased Risk in KD compared with Comparators (Yes, No, Unclear)	Bias (NOS: 0-9, JBI: 0-8)
Kitamura, 2009, ⁵⁴ Japan	RC	more cardiac interventions 114 KD patients	KD patients who underwent CABG for CAA	1 y	NR	114 (100)	19 y	Cardiac events (deaths, MI, syncope, angina, arrhythmias, cardiac interventions)	Cardiac events: 36 events in 33 patients. Event free rate was 87% at 5 y, 81% at 10 y, 67% (95%CI: 55% to 76%) at 20 y, and 60% (46–72) at 25 y	Unclear	NOS: 7
Suda, 2011, ⁴⁰ Japan	RC	76 KD patients	KD patients with giant aneurysms	Mean: 2.9 (2.9) y	NR	76 (100)	19 y (range: 2 mo–36 y)	MI, cardiac interventions (catheter/surgical)	MI: 12 (16%) patients; cardiac interventions: 46 (61%) patients. Cumulative coronary intervention rates: 28% at 5 y, 43% at 15 y, 59% at 25 y	Unclear	NOS: 6
Tsuda, 2011, ⁵⁵ Japan	RC	60 KD patients	KD patients with an acute MI from 1976–2007	1.4 (0.19–12) y	NR	Total: 60 (100); 50 (85%) had giant aneurysms	Median from initial MI: 16 y	HF, repeat MI, arrhythmia	HF: Of 45 patients alive, 45 were NYHA class I. Repeat MI: 13 patients experienced a second MI. Median interval between MI was 4 mo (range: 20 d–12.3 y). Arrhythmia: 6 patients (ventricular arrhythmia). Ventricular tachycardia-free survival rate after MI was 81% (95% CI, 68.9–89.2) at 10 y, 50% (35.6–64.5) at 20 y and 28.5% (15.4–46.5) at 25 y.	Unclear	NOS: 6
Holve, 2014, ⁴⁵ United States	RC	KD: 546 patients; general population comparators: 2218 patients	KD patients at least 1 y out from diagnosis and age of ≥15 y old at time of study	NR	Caucasian: 202 (37), Hispanic: 98 (17.9), Asian: 101 (18.5), Black: 71 (13), multiracial/	None: 436 (79.9); transient ectasia: 19 (3.5); persistent ectasia: 3 (0.5); transient	Mean 14.9 y	HTN, HLD, DM, MI, HF, arrhythmia	HTN: KD patients were more likely to have HTN than controls (5% vs 2%). HLD: 2% in KD group and 1% in controls. DM: 0% in KD group and 1% in controls. MI: 0.246/1000	HTN: Yes. HLD, DM, ACS, HF, arrhythmia: No	NOS: 8

SUPPLEMENTAL TABLE 11 Continued

Source	Study Design	Population	Selection Criteria	Median Age at KD Diagnosis	Ethnicity, n (%)	CAA, n (%)	Median Duration F/U (y)	Cardiac Outcome(s)	Results	Increased Risk in KD compared with Comparators (Yes, No, Unclear)	Bias (NOS: 0-9, JBI: 0-8)
Noto, 2014, Japan ⁴⁵	RC	58 KD patients	—	NR	other: 18 (3.3), unknown: 54 (9.9)	CAA: 23 (4.2); persistent CAA: 25 (4.6); unknown: 40 (7.3)	Mean 14.7 (5.5) y	MACE (MI, cardiac death, revascularization)	patient years in KD group and 0 in controls. HF: 0 in KD group and 1 in controls. Arrhythmia: 0 in KD group and 1 in controls. Any adverse outcome (includes mortality) occurring ≥ 15 y of age: No increase in events in KD group compared with controls (aHR: 0.81, 95% CI: 0.16-4.03). HTN was significantly associated with the outcome (HR: 7.1, 1.6-31.8)	MACE: Yes, KD patients with positive DSE had worse outcomes than negative DSE	NOS: 6
Tsuda, 2014, Japan ⁴⁶	RC	245 KD patients	KD patients with giant CAA	24 mo	NR	245 (100)	20 y	Cardiac events (death, cardiac interventions, MI, CABG, arrhythmia)	Cardiac interventions: 238 (97%) had catheterizations. CABG: CABG free rate at 30 y was 51% (95%CI, 45–60). Patients with bilateral giant CAA had lower CABG-free rate than unilateral giant CAA at 30 y. MI: 57(23%) had MIs, with	Cardiac events: Yes, KD patients with bilateral giant CAA had more events than unilateral giant CAA	NOS: 6

SUPPLEMENTAL TABLE 11 Continued

Source	Study Design	Population	Selection Criteria	Median Age at KD Diagnosis	Ethnicity, <i>n</i> (%)	CAA, <i>n</i> (%)	Median Duration F/U (y)	Cardiac Outcome(s)	Results	Increased Risk in KD compared with Comparators (Yes, No, Unclear)	Bias (NOS: 0-9, JBI: 0-8)
Hoshino, 2015, ³⁵ Japan	RC	20 KD patients	KD patients with systemic artery aneurysms from 1980–2013	6 mo (range: 1–20 mo)	NR	NR	18 y	Cardiac event (MI, CABG, cardiac interventions)	a median interval of 8 mo from diagnosis. MI free rate at 30 y was 74% (95%CI, 68–80). Patients with bilateral giant CAA had lower MI free rate than unilateral giant CAA at 30 y. Cardiac events: event free rate at 30 y was 36% (95%CI, 28–45). Patients with bilateral giant CAA had a lower event free rate than unilateral at 30 y (21% vs 59%). Cardiac events: 16 patients (80%) (7 MI, 8 CABG, 1 graft replacement). Cardiac event-free rate at 20 y: 29%	Unclear	NOS: 5
Lin, 2015, ⁴⁵ Taiwan	RC	1073 KD patients	—	1.59 y	NR	Total: 40.6% in acute fibrille stage, of which 196 (18.3) had CAA that persisted beyond 1 mo	5.63 y	Ischemia events (angina, MI, death)	Ischemia events: MI or death occurred in 14 patients (1%). Ischemia event-free rate at 10 y after KD diagnosis was 87.5% and at 20 y was 73.5%. For giant CAA patients (<i>n</i> = 27), ischemia event-free rate at 10 y was 63% and at 20 y was 36%	Unclear	NOS: 5
Bang, 2016, ³² Korea	RC	71 KD patients	KD patients with large CAA (≥6 mm)	Mean 4.6 (3.3) y	NR	71 (100)	Mean 12.5 (6.9) y	Cardiac intervention (PCI/CABG), MI	Cardiac interventions: 12 patients had PCI, at a median time of 5 y (0.8–12.3 y). 12 patients had CABG, at a median time of 8 y (2.1–14.7 y). There were no differences in cumulative coronary intervention rates between KD patients	Unclear	NOS: 5

SUPPLEMENTAL TABLE 11 Continued

Source	Study Design	Population	Selection Criteria	Median Age at KD Diagnosis	Ethnicity, n (%)	CAA, n (%)	Median Duration F/U (y)	Cardiac Outcome(s)	Results	Increased Risk in KD compared with Comparators (Yes, No, Unclear)	Bias (NOS: 0-9, JBI: 0-8)
Chih 2016, Taiwan ³⁶	RC	78 KD patients	KD patients with CAA \geq 4 mm	1.59 y	NR	Giant CAA (\geq 8mm): 27 (35); medium CAA (4–8 mm): 51 (65)	6.67 y	Ischemic disease (MI, CV-related death)	with 6–8 mm CAA vs >8 mm CAA. MI: 7 (9.9%) had MI, at a median time of 3 mo (0.5–31.2 mo) from KD diagnosis. Cumulative survival rate was 88.6%. Most MIs occurred in the first year after KD diagnosis Ischemic disease: giant CAA group (n = 27) had a 10-y survival of 66% and 20-y survival at 54%. Subgroup of patients with progressive coronary dilatation at least 2 mo after KD had lower freedom from ischemic events (OR: 4.81). Patients with progressive coronary dilatation at least 2 mo after KD had lower freedom from ischemic events (OR: 4.81). Patients with progressive coronary dilatation at least 2 mo after KD had lower ischemic-event free rate than KD patients with giant CAA (55% freedom at 5 y, 28% freedom at 10 y vs 74% freedom at 5 y, 59% freedom at 10 y)	Yes. KD patients with progression of CAA had more ischemic events than KD patients with giant CAA	NOS: 5
Tsuda, 2017, Japan ³⁷	RC	214 KD patients	KD patients with at least 1 CAA in a CA <100 d after KD onset	23 mo	NR	214 (100): small: 56; medium: 62; large: 96	16.8 y	Cardiac events (cardiac death, MI, cardiac procedures)	Cardiac events: occurred in 44 (21%) patients. Death occurred in 5, MI occurred in 20, and coronary procedures occurred in 36 patients. MI: 30-y MI-free survival in giant CAA group was 78% (95% CI, 70–87), and in medium CAA (\geq 6 mm	Yes. KD patients with giant CAA had more cardiac events than KD patients with medium CAA	NOS: 5

SUPPLEMENTAL TABLE 11 Continued

Source	Study Design	Population	Selection Criteria	Median Age at KD Diagnosis	Ethnicity, n (%)	CAA, n (%)	Median Duration F/U (y)	Cardiac Outcome(s)	Results	Increased Risk in KD compared with Comparators (Yes, No, Unclear)	Bias (NOS: 0-9, JBI: 0-8)
Dionne, 2017, Canada ¹²	RC	22 KD patients (10 KD patients post-PCI, 11 patients post-CABG, and 1 patient post-systemic thrombolysis)	KD patients with cardiac procedure (PCI/CABG/thrombolysis)	Mean 3.3 (3.3) y	NR	22 (100)	Mean time from CABG: 5.6 (4.1) y; Time from PCI: 6.5 (4.7) y	Cardiac re-intervention	Cardiac re-intervention: 6 patients required re-intervention, all previously treated by PCI. PCI group had significantly lower freedom from re-intervention/death versus CABG group	Yes. KD patients who had CABG had fewer reinterventions/death	NOS: 6
Wu, 2017, Taiwan ^{10,11}	RC	6690 KD patients	—	16 mo	NR	920 (14)	Mean: 10.45 (3.02) y	Major cardiac events (MI, cardiac procedures, death)	Major cardiac events: probability of major cardiac event was 1.9(0.3) at 15 y after KD or at age 15 y.	Unclear	NOS: 6
Miura, 2018, Japan ⁵⁰	RC	1006 KD patients	KD patients who received CA	1.8 y (IQR: 0.6–3.8)	NR	1006 (100): small: 134, medium: 425, large: 273, undassifiable: 174	6.4 y (IQR: 2.8–11.1)	Coronary events (coronary thrombosis, MI, coronary interventions), MADE (MI, coronary interventions, cardiac death)	Coronary events: 10-y event-free survival for small CAA: 100%, medium CAA: 96%, large CAA: 61%, MACE: 10-y event-free survival for small CAA: 100%, medium CAA: 98%, large CAA: 81%	Yes. KD patients with large CAA had more events	NOS: 5
Jeong, 2018, Korea ⁵¹	RC	20 KD patients	KD patients who underwent CABG	3 y (range: 0–7)	NR	20 (100)	Mean: 59.5 (46.5) mo	Repeat cardiac procedures	Repeat cardiac procedures: Freedom from revascularization at 10 y was 86.7% (SD: 8.9%)	Unclear	NOS: 5
Fukazawa, 2018, Japan ⁴⁷	RC	209 KD patients	KD patients with giant CAA (≥8 mm)	35 mo (range: 1–158 mo)	NR	209 (100)	Mean: 26 (13) mo	Cardiac events (cardiac death, MI, thrombolysis, CABG, catheter interventions)	Cardiac events: MI occurred in 32 (16%) with a median of 5 mo (range 0–85 mo) from diagnosis. 22 patients undergone CABG, with a median of 56 mo	Unclear	NOS: 6

SUPPLEMENTAL TABLE 11 Continued

Source	Study Design	Population	Selection Criteria	Median Age at KD Diagnosis	Ethnicity, n (%)	CAA, n (%)	Median Duration F/U (y)	Cardiac Outcome(s)	Results	Increased Risk in KD compared with Comparators (Yes, No, Unclear)	Bias (NOS: 0-9, JBI: 0-8)
Tadokoro, 2018, Japan ³⁹	RC	92 KD patients; single CABG; 53 patients; multiple CABG; 49 patients	KD patients who underwent CABG	NR, Mean patient age at surgery was 14.9 (10.4) y	NR	92 (100)	13 y (range: 1 mo–35 y)	Cardiac events (deaths, PCI, repeat CABG, MI, HF, ICD placement)	Cardiac events: 31 events in 25 patients (6 deaths, 12 PCI, 10 CABG, 1 MI, 1 CHF, 1 ICD placement). Cardiac event free survival at 15 y was 71% for single CABG group, 84% for multiple CABG group. Cardiac event free survival at 25 y was 45.2% at single CABG group and 68.5% at multiple CABG group. The reintervention-free survival rates were not significantly different between groups	No	NOS: 7
Yuan, 2020, China ⁴⁸	RC	38 KD patients	KD patients with giant CAA	<1 y old: 13 (34.2%); 1–5 y old: 19 (50.0%); ≥5 y old: 6 (15.8%)	NR	38 (100)	2.5 y	MACE (MI, cardiac death, cardiac interventions)	MACE: 8 (21.1%) experienced MACE with a median of 6.4 mo (range: 26 d–14.2 y) from diagnosis. MACE event free rates at 1 y was 86%, 2 y was 81%, and 5 y was 81%.	Unclear	NOS: 5

Risk of bias was evaluated with the NOS for case-control or cohort studies, and was evaluated with the JBI checklist for cross-sectional studies. AA, arterial aneurysms; ACS, acute coronary syndrome; aHR, adjusted hazard ratio; Apo, apolipoprotein; BAR, brachial artery reactivity; DaPW, brachial-ankle pulse wave velocity; BMI, body mass index; BP, blood pressure; BPs, bronchopulmonary sequestration; CA, coronary angiography; CC, case control; CHF, congestive heart failure; CI, confidence interval; cIMT, carotid intima-media thickness test; CS, cross-sectional; CV, cardiovascular; PC, prospective cohort; DBP, diastolic blood pressure; DM, diabetes mellitus; DSE, dobutamine stress echo ECHO, echocardiogram; EID, endothelium-independent dilatation; FMD, flow-mediated dilatation; HDL-C, high-density lipoprotein cholesterol; HF, heart failure; HLD, hyperlipidemia; HR, hazard ratio; HTN, hypertension; ICD, implantable cardioverter defibrillator; IQR, interquartile range; ITA, internal thoracic artery; LDL, low-density lipoprotein; LDL-C, low-density lipoprotein cholesterol; IMT, intima-media thickness; LVEF, low ventricular ejection fraction; NR, not reported; NYHA, New York Heart Association; OR, odds ratio; PCI, percutaneous coronary intervention; PWV, pulse wave velocity; RC, retrospective cohort; SBP, systolic blood pressure; SPECT, single-photon emission computerized tomography; SVG, saphenous vein graft; TC, total cholesterol; TG, triglycerides; —, not applicable.

SUPPLEMENTAL TABLE 12 KD Studies Which Evaluate Major Cardiovascular Event-Free Survival Over Time

Source	Population	Median Duration F/U (y)	Cardiac Event (CE) Definition	CE-Free Survival, % (95% CI)
Yoshikawa, 2000, Japan ³⁵	100 KD patients with obstructive CAA	Mean 6.7 (4.5) y	Postoperative MI, repeat CABG, catheter intervention	10 y: 70.5%
Yamauchi, 2004, Japan ⁴⁹	Total: 21 KD patients with CABG using arterial grafts. Group S (short duration from KD to CABG): 12 KD patients. Group L (long duration from KD to CABG): 9 KD patients.	Mean: group S: 5.5 (1.1) y; group L: 4.7 (1.1) y	HF, MI, valvular dysfunction	Over follow-up period: Group S 100%, group L 66.7%
Suda, 2009, Japan ⁴⁶	83 KD patients with giant CAA and on warfarin and aspirin ≥ 3 mo	Mean 5.8 (5.2) y	MI	6 mo: 92.7%; 1 y: 92.7%; 5 y: 91.2%; 10 y: 91.2%
Kitamura, 2009, Japan ³⁴	114 KD patients who underwent CABG	19 y	Cardiac deaths, MI, syncope, angina, arrhythmias, cardiac interventions	5 y: 87%; 10 y: 81%; 20 y: 67% (55–76); 25 y: 60% (46–72)
Suda, 2011, Japan ⁴⁰	76 KD patients with giant CAA	19 y (range: 2 mo–36 y)	Cardiac interventions (catheter/surgical)	5 y: 72%; 15 y: 57%; 25 y: 41%
Tsuda, 2011, Japan ³⁵	60 KD patients with an acute MI from 1976–2007	Median from initial MI: 16 y	Ventricular tachycardia post MI	10 y: 81% (68–89); 20 y: 50% (36–64); 25 y: 28.5% (15–46)
Tsuda, 2014, Japan ³⁶	245 KD patients with giant CAA	20 y	Cardiac death, cardiac interventions, MI, CABG, arrhythmia	30 y: 36% (28–45); CABG-free rate: 30 y: 51% (43–60)
Hoshino, 2015, Japan ⁵³	20 KD patients with systemic artery aneurysms	18 y	MI, CABG, cardiac interventions	20 y: 29%
Lin, 2015, Taiwan ⁴⁵	1073 KD patients	5.63 y	Angina, MI, cardiac death	10 y: 87.5%; 20 y: 73.5%; giant CAA patients: 10 y: 63%; 20 y: 36%
Bang, 2016, Korea ⁵²	71 KD patients with large CAA (≥ 6 mm)	Mean 12.5 (6.9) years	MI	Over follow-up period: 89.6%
Chih 2016, Taiwan ³⁸	78 KD patients with CAA ≥ 4 mm	6.67 y	MI, cardiac death	10 y: 66%; 20 y: 54%
Tsuda, 2017, Japan ³⁷	214 KD patients with at least 1 CAA in a CA < 100 d after KD onset	16.8 y	MI, cardiac procedures	MI: 30 y; giant CAA: 78% (70–87); medium CAA (≥ 6 mm and < 8 mm): 96% (86–99); coronary procedures: 30 y; giant CAA: 59% (48–69); medium CAA: 85% (72–93)
Miura, 2018, Japan ⁵⁰	1006 KD patients with CAA	6.4 y (IQR: 2.8–11.1)	MI, coronary interventions, cardiac death	10 y: small CAA: 100%; medium CAA: 98% Large CAA: 81%
Jeong, 2018, Korea ⁵¹	20 KD patients with CABG	Mean: 59.5 (48.5) mo	Repeat cardiac procedures	10 y: 86.7%
Fukazawa, 2018, Japan ⁴⁷	209 KD patients with giant CAA (≥ 8 mm)	Mean: 26 (13) mo	Cardiac death, MI, thrombolysis, CABG, catheter interventions	1 y: 80%; 5 y: 72%; 10 y: 68%
Tadokoro, 2019, Japan ³⁹	92 KD patients with CABG; single CABG: 53 patients; multiple CABG: 49 patients	13 y (range: 1 mo–35 y)	Cardiac deaths, PCI, repeat CABG, MI, HF, ICD placement	15 y: single CABG: 71.0%; multiple CABG: 84.0%; 25 y: single CABG: 45.2%; multiple CABG: 68.5%
Yuan, 2020, China ⁴⁸	38 KD patients with giant CAA	2.5 y	MI, cardiac death, cardiac interventions	1 y: 86%; 2 y: 81%; 5 y: 81%

CA, coronary angiography; CE, cardiac event; F/U, follow-up; HF, heart failure; ICD, implantable cardioverter defibrillator; IQR, interquartile range; PCI, percutaneous coronary intervention.

SUPPLEMENTAL TABLE 13 KD Studies Evaluating Chronic Cardiovascular Diseases, including Hypertension (HTN), Hyperlipidemia (HLD), and Early Atherosclerosis

Source	Population	Median Duration F/U (y)	Results	Increased Risk in KD Compared With Selected Comparators?
Novelli, 1984, United Kingdom ⁵⁶	18 KD patients	Mean 12.3 mo	HTN: 8 (44%), 6 patients were mild without treatment	Unclear
Newburger, 1989, Japan ⁵⁷	105 KD patients, of which 46 KD patients were compared with parents.	NR. 30 patients had lipid profiles measured at >3 y	HLD: Total cholesterol at time period 1 (<10 d from KD) was significantly lower than all periods. HDL was depressed <10 d from KD and then increased significantly with time, with the highest at period 6 (>3 y from KD), but when adjusted, KD patients had HDL values that were significantly lower than normal. Subgroup compared with parents: Adjusted total cholesterol not significantly different from parent(s), but had significantly lower HDL than parents	Yes
Noto, 2001, Japan ⁵⁹	20 KD patients with CAA and 20 healthy controls (age and sex matched)	Mean 9.8 (4) y	HTN: No significant difference in systolic or diastolic BPs between groups. HLD: No significant difference in TG, Total cholesterol, HDL between groups. TG, Total cholesterol, LDL, HDL all within normal limits in KD. IMT: Significantly higher in KD than in controls (4.11 [0.86] mm vs 2.94 [0.91] mm)	HLD: No. HTN: No. IMT: Yes
Niboshi, 2007, Japan ¹⁰⁶	Group A: 9 KD patients with CAA; group B: 6 KD patients with transient CAA; group C: 20 KD patients without CAA; group D: 36 non-KD Controls	Mean 24.1 y	HLD: No difference in TG, HDL, LDL, TC/HDL Ratio between KD groups and controls. Endothelial dysfunction: %FMD and FMD/EID ratio was markedly reduced in KD group compared with controls. FMD was reduced more in group A compared with group C. Arterial stiffness: baPWV values were significantly increased in male KD groups compared with male controls	HTN, HLD: No. Endothelial dysfunction: Yes
Ou, 2009, Taiwan ⁶³	Group 1: 55 KD patients with persistent CAA; group 2: 64 KD patients without CAA	Mean: group 1: 2.46 y (range 0.71–4.62); group 2: 2.52 y (1.12–4.92)	HTN: groups did not differ significantly between sBP and dBP. HLD: no difference in LDL-C, TG, total cholesterol. HDL levels were significantly different: group 1: 42.5 (SD: 10.23) mg/dL, group 2: 44.23 (10.91) mg/dL	HTN: No. HLD: Unclear

SUPPLEMENTAL TABLE 13 Continued

Source	Population	Median Duration F/U (y)	Results	Increased Risk in KD Compared With Selected Comparators?
Cho, 2013, Korea ⁶⁴	Group A: 19 KD patients with persistent/regressed CAA; group B: 49 KD patients without CAA; group C: 30 age- and sex-matched non-KD patients	Mean: group A: 68.95 (31) mo; group B: 52.29 (27) mo	HTN: no difference among the 3 groups. HLD: levels of TC, LDL, Apo-B were higher in group A than C. No difference between group A and B. No difference between Apo-A, HDL-C, and TG between groups. Arterial stiffness: PWV of KD patients were significantly higher in KD groups than group C	HTN: No. HLD: Unclear. Arterial stiffness: Yes
Ishikawa, 2013, Japan ⁶⁰	Group 1: 9 KD patients with CAA; group 2: 15 KD patients without CAA; group 3: 22 age- and sex-matched non-KD patients	Group 1: 4.0 y (range 3.2–4.1); group 2: 2.0 y (range 1.0–3.4)	HTN: No significant difference. HLD: No significant difference in TC, HDL, LDL, TC/HDL, TG. Endothelial Dysfunction: %FMD was significantly lower in Group 1 compared with group 2 and group 3. %FMD in group 2 was significantly lower than group 3	HTN, HLD: No. Endothelial Dysfunction: Yes
Holve, 2014, United States ⁴⁵	KD: 546 patients. General population comparators: 2218 patients	Mean 14.9 y	HTN: KD patients were more likely to have HTN than controls (3.1% vs 1.7%). HLD: 2.2% in KD group and 1.4% in controls	HTN: Yes. HLD: No
Parihar, 2017, India ¹¹⁰	20 KD patients, 20 age- and sex-matched controls	Mean: 53.7 mo	Arterial stiffness: cIMT was similar in cases and controls and there was no significant difference in carotid artery stiffness index	No
Gopalan, 2018, India ¹⁷	27 KD patients; 27 age- and sex- matched comparators or closest sibling	Mean: 83.77 (14.12) mo	HLD: abnormal lipid profile was seen in 7 patients. There was no statistically significant difference in cholesterol, HDL, LDL, TG levels between KD patients with and without CAA at diagnosis. Arterial stiffness: mean cIMT was higher in KD patients than controls	HLD: Unclear. Arterial stiffness: Yes
Cheung, 2004, Hong Kong ⁵⁸	Group 1: 37 KD patients with CAA; group 2: 29 KD patients without CAA; group 3: 36 healthy aged-matched children	Mean: group 1: 7.8 (3.7) y; group 2: 6.2 (2.4) y	HLD: HDL was significantly lower in group 1 compared to group 2 or group 3. ApoA-I levels were significantly lower in group 1 compared to group 2 or group 3. There were no significant differences in TC or TG. HTN: No difference between the 3 groups. Arterial stiffness: PWV of group 1 was significantly higher than group 3. PWV of group 2 was significantly higher than group 3. There	HLD: Yes. HTN: No. Arterial stiffness: Yes

SUPPLEMENTAL TABLE 13 Continued

Source	Population	Median Duration F/U (y)	Results	Increased Risk in KD Compared With Selected Comparators?
Cheung, 2006, Hong Kong ⁶⁵	Group 1: 26 KD patients with CAA; group 2: 24 KD patients without CAA; group 3: 22 healthy age-matched children	Mean: group 1: 7.4 (3.5) y; group 2: 5.8 (2.1) y	was no significant difference between groups 1 and 2 Arterial stiffness: stiffness index and brachioradial PWV differed significantly between groups, with group 1 having a higher stiffness index than group 2 and group 2 having a higher stiffness index than group 3. group 1 had faster PWV than group 3. cIMT differed significantly between the 3 groups with group 1 and 2 having greater cIMT than group 3. cIMT correlated with LDL, stiffness index, and PWV. HTN: No difference between groups. HLD: No difference in TC or HDL between groups. Group 1 had higher LDL compared to groups 2 and 3	Arterial stiffness: Yes. HTN: No. HLD: Unclear
McCordle, 2007, Canada ⁶¹	52 KD patients and 60 healthy controls between 10–20 y old	Mean: 11.2 (0.37) y	HTN: KD patients had lower SBP. No significant difference in 24-h ambulatory BP monitoring. HLD: KD patients had lower levels of ApoA-I. There was no significant difference in the remaining lipid profiles. Arterial stiffness: no significant difference. No relationship noted with presence/absence of CAA	HTN: No. HLD: No. Arterial stiffness: No
Chen, 2017, Australia ¹¹¹	Australia cohort: 57 KD patients and 59 age and sex-matched controls. Singapore cohort: 78 KD patients and 78 age- and sex-matched controls	Mean: Australia: 11.8 (5.9) y; Singapore: 6.9 (4.0) y	Retinal microvasculature: KD patients from Singapore had larger retinal venules than controls. KD patients with CAA from Australia had larger retinal venules than controls but not significant. Retinal venular caliber increases were not associated with age, sex, time since KD, mean BP, or BMI	Unclear

Apo, apolipoprotein; baPWV, brachial-ankle pulse wave velocity; BMI, body mass index; BP, blood pressure; dBp, diastolic blood pressure; cIMT, carotid intima-media thickness test; EID, endothelium-independent dilatation; FMD, flow-mediated dilatation; F/U, follow-up; HDL-C, high-density lipoprotein cholesterol; HLD, hyperlipidemia; HTN, hypertension; IMT, intima-media thickness; LDL, low-density lipoprotein; NR, not reported; PWV, pulse wave velocity; SBP, spontaneous bacterial peritonitis; SBP, systolic blood pressure; TC, total cholesterol; TG, triglycerides

SUPPLEMENTAL TABLE 14 Studies Evaluating Noncardiovascular Outcomes in the KD Population

Source	Study Design	Recruitment Setting	Population	Median (IQR) Age at KD Diagnosis	Ethnicity, n (%)	Mean (SD)/Median (IQR) Duration F/U (y)	Analysis	Results	Increased Risk in KD population	Bias (NOS: 0-9, JBI: 0-8)
Allergic diseases (asthma, atopic dermatitis (AD), allergic rhinitis (AR), allergies, allergic conjunctivitis)										
Brosius, 1988, ⁷³ United States ⁷³	CC	Hospital	KD: 83; healthy controls: 83	NR	Caucasian: 66 (80), Black: 6 (7), Asian: 4 (5), Hispanic: 3 (2), mixed: 4 (6)	Mean ^a : 1.7 (0.2) y	Univariable analysis	AD: OR 9.0 ^b ; eczema: OR 2.5	Yes	NOS: 5
Liew, 2011, Singapore ⁶⁹	CS	Hospital	KD: 93; healthy siblings: 93	14 mo	Chinese: 72 (78), Malaysian: 18 (19), Indian: 1 (1), other: 2 (2)	NR. Minimum 1 y out from KD diagnosis	Multivariable logistic regression	AR: OR 2.40. No difference in 'any allergies', asthma, eczema, or allergic conjunctivitis	Yes	JBI: 6
Hwang, 2013, Taiwan ⁷²	CC	Population-based	KD: 253; general population: 800	Mean: 3 (1.3) y	NR	NR. Maximum 13 y of observation.	Multivariable logistic regression	Any atopic disease ^c : aOR 1.61 (95% CI, 1.15-2.26). Age- and sex-stratification showed males between 1-5 y old at highest risk (OR 1.95; 95% CI, 1.20-3.17)	Yes	NOS: 7
Kuo, 2013, Taiwan ⁷⁰	RC	Population-based	KD: 253; general Population: 1012	Mean: 1.61(1.20) y	NR	NR. Maximum 5 y of observation	Survival analysis, multivariable Cox regression analysis	Increased incidence rate of allergic disease (asthma and AR) in KD: 184.66 per 1000 PY. Asthma: aHR 1.51 (95% CI: 1.17-1.95). AR:	Yes	NOS: 8

SUPPLEMENTAL TABLE 14 Continued

Source	Study Design	Recruitment Setting	Population	Median (IQR) Age at KD Diagnosis	Ethnicity, n (%)	Mean (SD)/Median (IQR) Duration F/U (y)	Analysis	Results	Increased Risk in KD population	Bias (NOS: 0-9, JBI: 0-8)
Tsai, 2013, Taiwan ⁷¹	CC	Population-based	KD: 7072; general population: 27 265	NR	NR	NR. range of 6–13 y after KD diagnosis, maximum 13 y of observation	Multivariable logistic regression	aHR 1.30 (95% CI: 1.04–1.62) AR: OR 1.28 (95% CI, 1.20–1.37). Asthma, AD, urticaria: not significant in multivariable models	Yes	NOS: 8
Baker, 2003, United States ^{4,102}	CS	Hospital	KD: 110	3.1 y (range: 0.2–12)	White: 77%, Black: 6%, Hispanic: 7%, Asian: 6%, other: 2%, unknown: 1%	6.5 y (range: 0.2–15.8)	Univariable analysis	Compared with a US population sample, KD patients reported a higher prevalence of chronic allergies and sinus trouble (26.9% vs 17.6%)	Yes	JBI: 6
Cognitive impairment Wang, 2018, Taiwan ¹⁰³	RC	Hospital and population-based	Clinical cohort: 168 KD patients, 81 healthy controls; population-based: 4286 KD patients, 50 038 comparators	4–7 y group: mean 23.12 (17.51) mo; >7 y group: 21.14 (16.51) mo	NR	3 y	Multivariable Cox regression analysis	Clinical study: no difference in developmental index or cognitive function between KD patients and controls. Population based study: no difference in the prevalence of intellectual disability. KD was not associated with increased risk of intellectual	No	NOS: 7

SUPPLEMENTAL TABLE 14 Continued

Source	Study Design	Recruitment Setting	Population	Median (IQR) Age at KD Diagnosis	Ethnicity, n (%)	Mean (SD)/Median (IQR) Duration F/U (y)	Analysis	Results	Increased Risk in KD population	Bias (NOS: 0-9, JBI: 0-8)
Attention deficit hyperactivity disorder (ADHD) Kuo, 2015, Taiwan ⁷⁰	RC	Population-based	KD: 651; general population: 3255	NR	NR	Mean: 4.93 y	Survival analysis, multivariable Cox regression analysis	disability (aHR = 0.87; 95% CI: 0.68–1.11) After adjusting for confounders (ie, sex, age, relevant diseases like allergic diseases or meningitis), KD was not associated with increased risk of ADHD (aHR = 1.59; 95% CI, 0.96–2.62)	No	NOS: 8
Cancer Yu, 2016, Taiwan ²⁴	RC	Population-based	KD: 3469	Mean 3.02 (3.81)	NR	NR. Maximum 15 y of observation	Incidence rates	Incidence rate: 34.7 per 100 000 person-years. SIR: 2.88 (95% CI, 1.60–5.20). KD patients were higher in younger patients (<20 y old) at SIR 3.13 (1.73–5.65), and patients with longer follow-up from diagnosis (>6 y) at SIR 5.34 (2.01–14.24)	Yes	NOS: 6
Physical and psychosocial well-being										

SUPPLEMENTAL TABLE 14 Continued

Source	Study Design	Recruitment Setting	Population	Median (IQR) Age at KD Diagnosis	Ethnicity, n (%)	Mean (SD)/Median (IQR) Duration F/U (y)	Analysis	Results	Increased Risk in KD population	Bias (NOS: 0-9, JBI: 0-8)
Baker, 2003, United States ¹⁰²	CS	Hospital	KD: 110	3.1 y (range: 0.2-12)	White: 77%, Black: 6%, Hispanic: 7%, Asian: 6%, other: 2%, unknown: 1%	6.5 y (range: 0.2-15.8)	Univariable analysis	Physical summary score: KD patients with giant aneurysms had a significantly lower score than the US population (47.3 [9.7] vs 53 [8.8]). No differences in psychosocial summary score in KD versus general population. KD patients reported a higher prevalence of bone and joint problems (11.1% vs 3.4%) and anxiety (13.9% vs 5.2%)	Yes	JBI: 6
Exercise performance Paridon 1995, United States ¹⁰⁵	CS	Hospital	KD: 46; patients, grouped by presence or absence of CAA	Mean 4.0 (2.0) y	NR	Range of 5-7 y from diagnosis	Univariable analysis	No difference in maximal HR, maximal $\dot{V}O_2$, maximal respiratory exchange ratio in 3 groups	No	JBI: 8
Obstetric outcomes Tsuda, 2006, Taiwan ¹⁰⁶	CS	Hospital	KD: 30; patients with CAA	NR	NR	Median maternal age at delivery: 27 y (range 18-35 y)	Univariable analysis	None of the patients had a cardiac event during pregnancy. 2	No	JBI: 3

SUPPLEMENTAL TABLE 14 Continued

Source	Study Design	Recruitment Setting	Population	Median (IQR) Age at KD Diagnosis	Ethnicity, n (%)	Mean (SD)/Median (IQR) Duration F/U (y)	Analysis	Results	Increased Risk in KD population	Bias (NOS: 0-9, JBI: 0-8)
Respiratory tract infections (RTI) Lei, 2018, Taiwan ¹⁰⁷	RC	Population-based	KD: 4973. General population: 19 683	73.8% were <2 y old, 26.2% were 2-6 y old	NR	Observed up to 1 y from diagnosis	Survival analysis, multivariable Cox regression analysis	patients had obstetric complications (PPROM, hemorrhage) Cumulative incidence of RTI-related hospitalizations was lower in the KD cohort than in the non-KD cohort. aHR or RTI-related hospitalizations: 0.75 (95% CI, 0.66-0.85)	No	NOS: 8

Risk of bias was evaluated with the NOS for case-control or cohort studies, and was evaluated with the JBI checklist for cross-sectional studies. AD, atopic dermatitis; ADHD, attention-deficit/hyperactivity disorder; aHR, adjusted hazard ratio; AR, allergic rhinitis; CI, confidence interval; CS, cross-sectional; HR, heart rate; maximal vO2, maximum rate of oxygen consumption; NR, not reported; OR, odds ratio; PPRM, preterm premature rupture of membranes; PY, person-years; RC, retrospective cohort; RTI, respiratory tract infection; SIR, standardized incidence ratio.

^aMean provided with SD.

^bAtopic dermatitis classified as a patient with eczema and a personal or family history (first-degree relative) of inhalant allergies (ie, allergic rhinitis) or asthma.

^cAtopic disease: atopic dermatitis, allergic rhinitis, asthma.

^dBaker et al is described twice in this chart because they include numerous noncardiac outcomes, including allergic disease.

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