Access To Care: HCM Roundtable 2021

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Disclosures

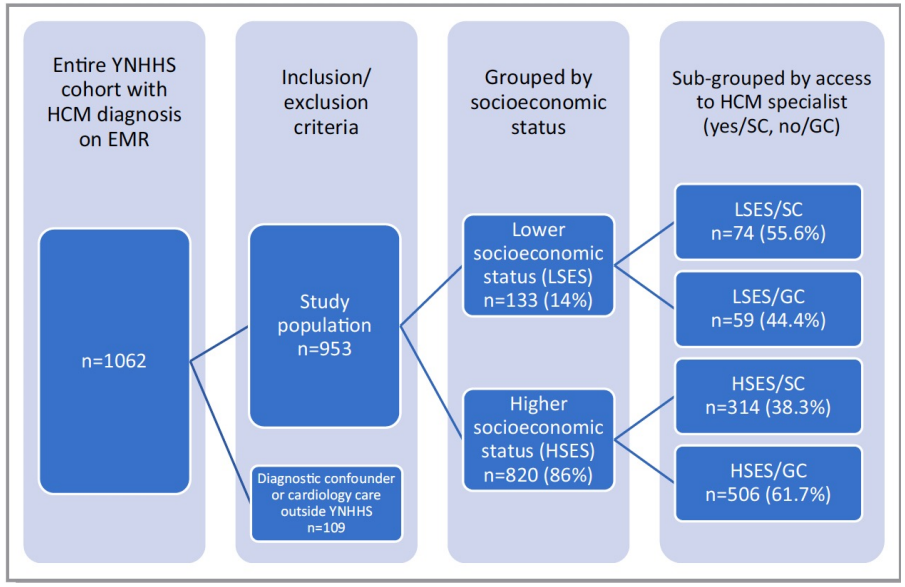
Myokardia: Grants, Steering Committee EXPLORER

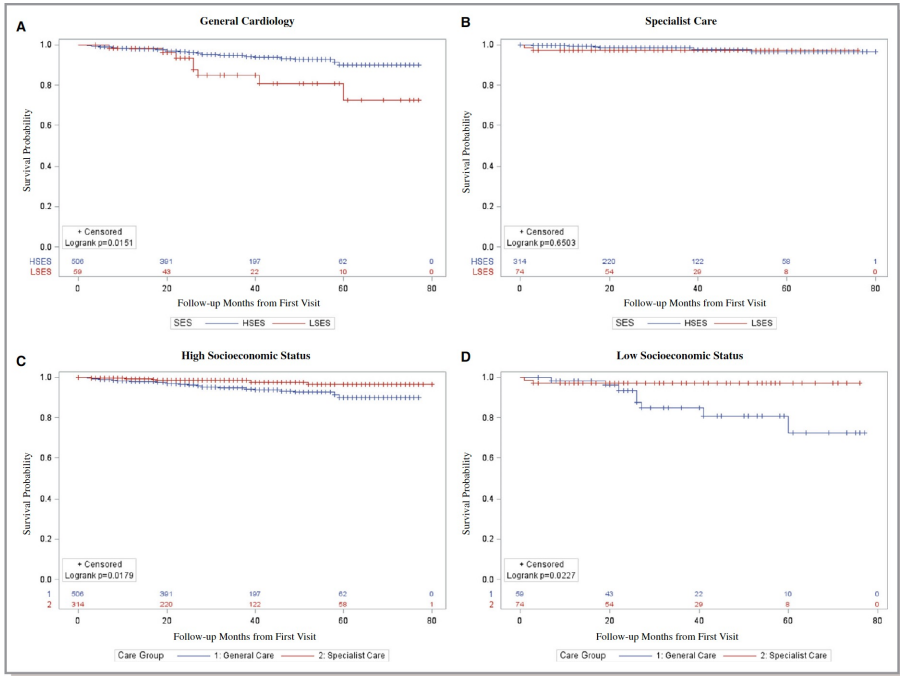
Cytkonetics: Consulting

Propria LLC: Patented technology for pre-clinical diagnostics in HCM, patent ownership (self) and company ownership (spouse)

Access to specialty care is unequal and changes outcome

Thomas and Jacoby, J Am Heart Assoc. 2020





Does resource use explain differential outcome?

Table 4. Both LSES and HSES Patients Were at Hi
of Being Hospitalized If They Received Specialty Ca

	HR (95% CI)	P
LSES		
Specialist care	3.28 (1.11–9.73)	0.
General cardiology	1	
HSES		
Specialist care	2.19 (1.40–3.40)	0.
General cardiology	1	
Interaction (moderation), HRR	1.50 (0.47–4.85)	0.
Age, y	1.02 (2.73–2.79)	0.
Male	0.66 (1.56–2.68)	0.
Race		
Black (vs white)	0.99 (1.69–6.54)	0.
Other (vs white)	1.38 (1.89–19.47)	0.
Unknown (vs white)	1.12 (1.36–55.15)	0.
Ethnicity		
Hispanic (vs non-Hispanic)	1.16 (1.68–13.71)	0.
Unknown (vs non-Hispanic)	0.12 (1.02–2.10)	0.
DM	1.08 (1.86–6.48)	0.
CAD	1.75 (2.82–19.38)	0.036*

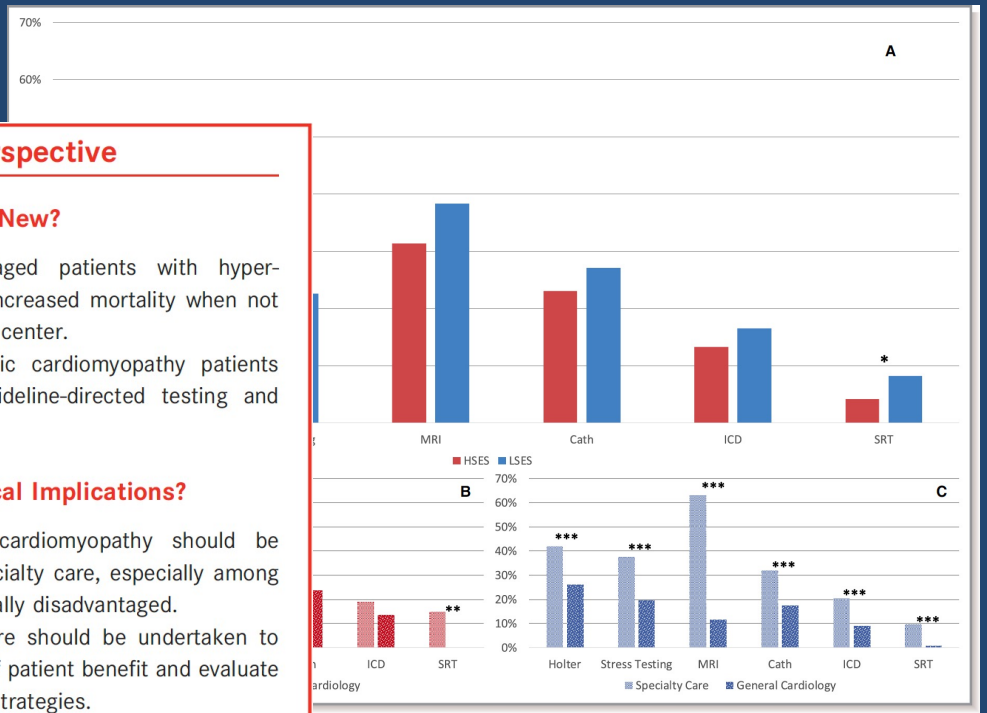
Clinical Perspective

What Is New?

- Socioeconomically disadvantaged patients with hypertrophic cardiomyopathy had increased mortality when not involved in care at a specialty center.
- Specialty care of hypertrophic cardiomyopathy patients leads to more-consistent guideline-directed testing and treatments.

What Are the Clinical Implications?

- Patients with hypertrophic cardiomyopathy should be considered for referral to specialty care, especially among those who are socioeconomically disadvantaged.
- Further study of specialty care should be undertaken to better assess for the drivers of patient benefit and evaluate for improved implementation strategies.



mended testing for hypertrophic cardiomyopathy (HCM) comparing high and low socioeconomic groups as well as specialty care and general cardiology subgroups. **A**, Comparison of HSES and LSES groups. **B**, Comparison of HCM patients with LSES between specialty care and general cardiology subgroups. **C**, Comparison of HCM patients with HSES between specialty care and general cardiology subgroups. *P<0.05; **P<0.01; ***P<0.001. Septal reduction therapy includes alcohol ablation procedures and myectomies. Cath indicates cardiac catheterization; HSES, high socioeconomic status; ICD, implantable cardiac defibrillator; LSES, low socioeconomic status; MRI, magnetic resonance imaging; SRT, septal reduction therapy.

Similarly, disease expression and outcomes diverge by race

Eberly and Lakdawala, JAMA Cardiol. 2020

Table 1. Characteristics of Hypertrophic Cardiomyopathy Popu
Specialty Care

	Higher Socioeconomic Status (HSES)		
	SC	GC	Total
n (%)	314 (38)	506 (62)	820
Age, y (SD)	54.67 (15.6)	65.2 (17.3)	61.1 (17.4)*
Sex, n (%)			
Male	199 (63)	273 (54)	472 (58)*
Female	115 (37)	223 (46)	348 (42)*
Race, n (%)			
White	251 (80)	392 (77)	643 (78)
Black	31 (10)	48 (10)	79 (10)
Asian	3 (1)	5 (1)	8 (1)
Other/unknown	29 (9)	61 (12)	90 (11)
Comorbidities, n (%)			
CAD	33 (11)	75 (15)	108 (13)
DM	31 (10)	87 (17)	118 (14)*
Hypertension	159 (51)	313 (62)	472 (56)*

Key Points

Question Is race associated with differential disease expression, inequitable care provision, or disparate clinical outcomes among patients with hypertrophic cardiomyopathy?

Findings In this cohort study of 2467 patients with cardiomyopathy, compared with white patients, black patients with hypertrophic cardiomyopathy were diagnosed at a younger age, were less likely to have sarcomere mutations, and had worse symptoms. Inequities in health care access and delivery were associated with race, with lower rates of genetic testing and invasive septal reduction therapy among black patients with hypertrophic cardiomyopathy.

Meaning The findings suggest that racial differences in disease expression and adverse clinical outcomes exist between black and white patients with hypertrophic cardiomyopathy and that these differences may be associated with inequities in clinical care provision.

mes Among Black and White Patients

	No. (%)		
	Black Patients (n = 205)	White Patients (n = 2262)	*P* Value
	30 (14.6)	521 (23.0)	.007
			.03
	2 (6.7)	59 (11.3)	NA
	28 (93.3)	447 (85.8)	NA
	0	15 (2.9)	NA
	68 (33.2)	724 (32.0)	.79
	13 (6.3)	119 (5.3)	.43
	0	16 (0.7)	.45
	11 (5.4)	74 (3.3)	.17
	6 (2.9)	92 (4.1)	.54
	35 (17.1)	608 (26.9)	<.001
	10 (4.9)	80 (3.5)	.43
	2 (1.0)	56 (2.5)	.17
	51 (24.8)	440 (19.5)	.050
	11 (5.4)	159 (7.0)	.37
	16 (7.8)	157 (6.9)	.75
	91 (44.4)	1003 (44.3)	>.99

mponents in Black vs White Patients

Referrals and access

Outcomes studies and referrals

Comparison of Clinical Features in Blacks Versus Whites With Hypertrophic Cardiomyopathy

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To date, there has not been a large systematic examination of the hypertrophic cardiomyopathy (HC) phenotype in blacks versus whites. In this study, we investigate differences in presentation of HC between blacks and whites. We included 441 consecutive patients with HC seen at the Johns Hopkins HC clinic in the period from February 2005 to June 2012. We compared 76 blacks for clinical presentation, electrocardiogram, exercise capacity, left ventricular morphology, and hemodynamics by echocardiography to 365 whites. Black patients with HC more often presented with abnormal electrocardiogram (93% vs 80%, p = 0.009), driven by a significant difference in repolarization abnormalities (79% vs 56%, p <0.001). Apical hypertrophy was more common in blacks (26% vs 9%, p <0.001); however, blacks had less severe systolic anterior movement of the mitral valve and had significantly lower left ventricular outflow tract gradients at rest (9 mm Hg; interquartile range [IQR] 7 to 19 vs 16 mm Hg; IQR 8 to 40, p <0.001) and during provocation (36 mm Hg; IQR 16 to 77 vs 59 mm Hg; IQR 26 to 110, p = 0.002). Despite the nonobstructive pathophysiology, blacks had lower exercise capacity (adjusted difference 1.45 metabolic equivalents [0.45 to 2.45], p = 0.005). In conclusion, blacks have an HC phenotype characterized by lower prevalence of the well-recognized echocardiographic features of HC such as systolic anterior movement of the mitral valve and left ventricular outflow tract obstruction and display worse exercise capacity. (Am J Cardiol 2016;117:1815–1820)

RESEARCH LETTER

Association Between Race and Clinical Profile of Patients Referred for Hypertrophic Cardiomyopathy

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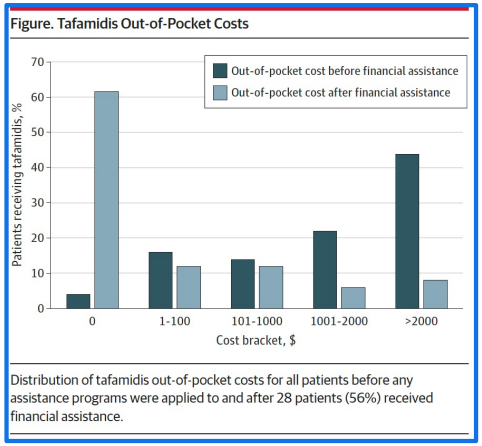
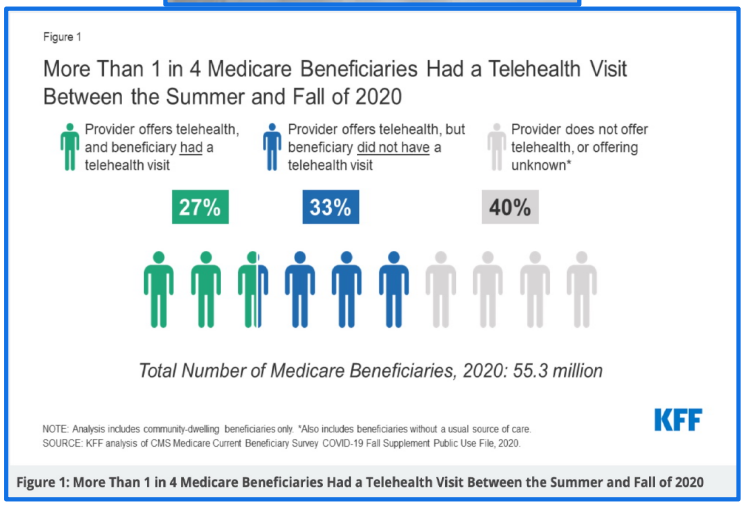
Hypertrophic cardiomyopathy (HCM) is a genetic heart disease that affects patients globally, with an estimated prevalence of at least 1:500.[1] Despite significant progress, little is known regarding the specific role of race in clinical course and presentation.[2-4] However, excess HCM-related sudden deaths in black male athletes justify the current initiative exploring racial differences in disease progression and referral patterns for patients with HCM.[2-5]

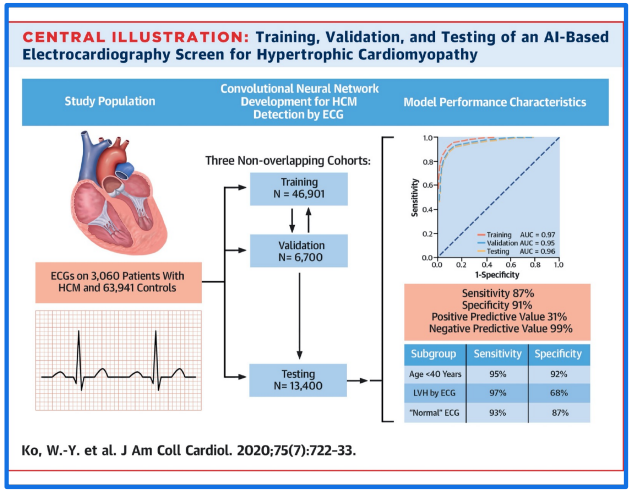
Of 1900 patients referred to HCM programs at Tufts and Morristown Medical Centers from 2004 to 2017, 90 consecutive black patients were identified and followed for 3±3 years. Race was self-reported, including African-American or those of recent African or Caribbean descent. Patients met diagnostic criteria for HCM with unexplained hypertrophy and nondilated left ventricle. Patients were 47±17 years of age at initial evaluation, 59% male, and matched to 270 consecutive white patients at the same centers over the same time period, 3:1 for age and sex. This study was reviewed and approved by the Tufts Institutional Review Committee in accordance with journal guidelines.

The impact of innovative therapies and technology are likely to be complex



Masri and Heitner, JAMA Card 2020





Ko and Noseworthy, JACC 2020

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Under-representation has adverse consequences

Clinical trials and gene testing

	Mavacamten group (n=123)	Placebo group (n=128)
Age, years	58·5 (12·2)	58·5 (11·8)
Sex		
Women	57 (46%)	45 (35%)
Men	66 (54%)	83 (65%)
Race		
White	115 (93%)	114 (89%)
Black or African American	1 (1%)	5 (4%)
Native American or Alaskan Native	0	1 (1%)
Asian	4 (3%)	2 (2%)
Unknown	3 (2%)	6 (5%)
Region		
USA	53 (43%)	55 (43%)
Spain	17 (14%)	16 (13%)
Poland	16 (13%)	16 (13%)
Other*	37 (30%)	41 (32%)

Table 1, EXPLORER HCM
Olivotto and Jacoby, Lancet 2020

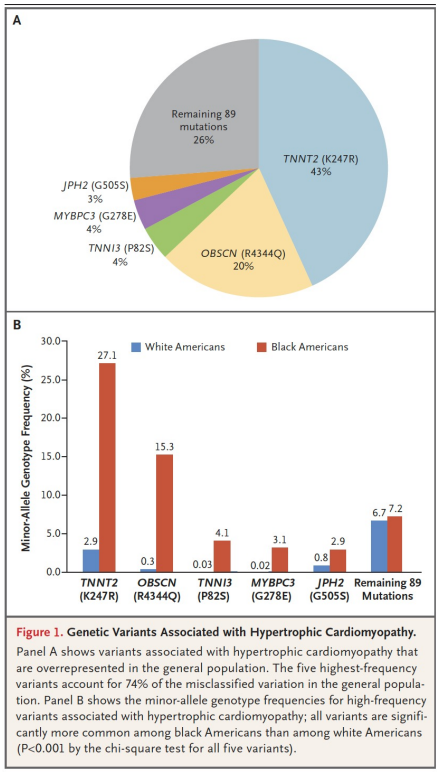

Figure 1. Genetic Variants Associated with Hypertrophic Cardiomyopathy.
Panel A shows variants associated with hypertrophic cardiomyopathy that are overrepresented in the general population. The five highest-frequency variants account for 74% of the misclassified variation in the general population. Panel B shows the minor-allele genotype frequencies for high-frequency variants associated with hypertrophic cardiomyopathy; all variants are significantly more common among black Americans than among white Americans ($P<0.001$ by the chi-square test for all five variants).

Manrai and Kohane, NEJM 2016

Goals and associated process metrics are required

ACGME

Improve access to specialty care for all

Improve representation of minority patients in clinical trials and outcome studies

Educate providers and develop tools to diagnose patients

Train next generation of providers

Questions

Thank you

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