

2023 CASCADE FH[®] Registry

National Lipid Association
Atlanta, GA | June 3, 2023

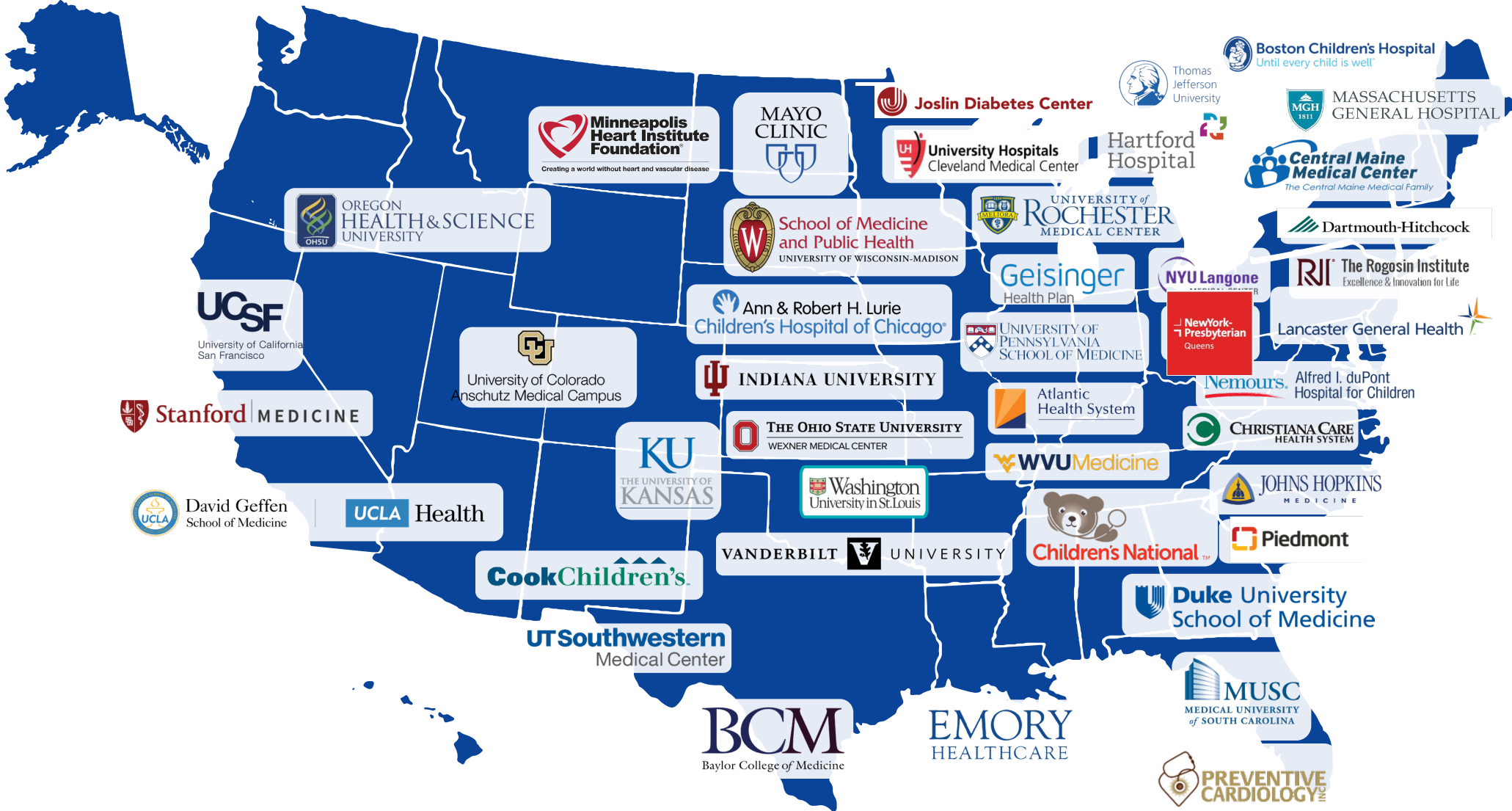
Disclosures- none to report

Objectives

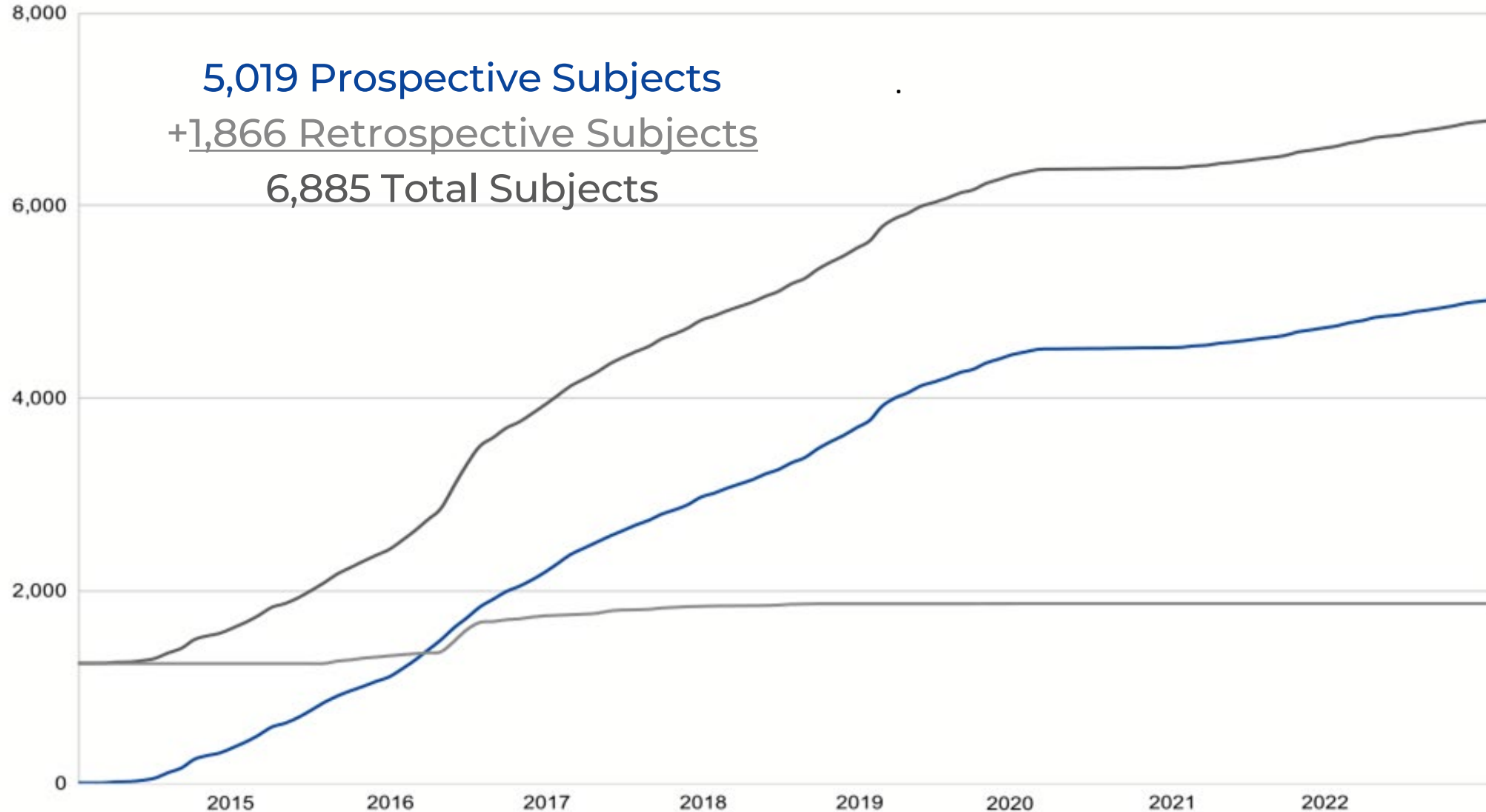
At the end of this presentation participants will be able to:

1. Describe characteristics of the CASCADE FH Registry population
2. Discuss the need for using multiple lipid lowering medications for the Treatment of FH
3. Recognize the Family Heart Foundation as a patient resource (familyheart.org)

CASCADE FH Network



CASCADE FH Registry Enrollment



Family Heart CASCADE FH Registry

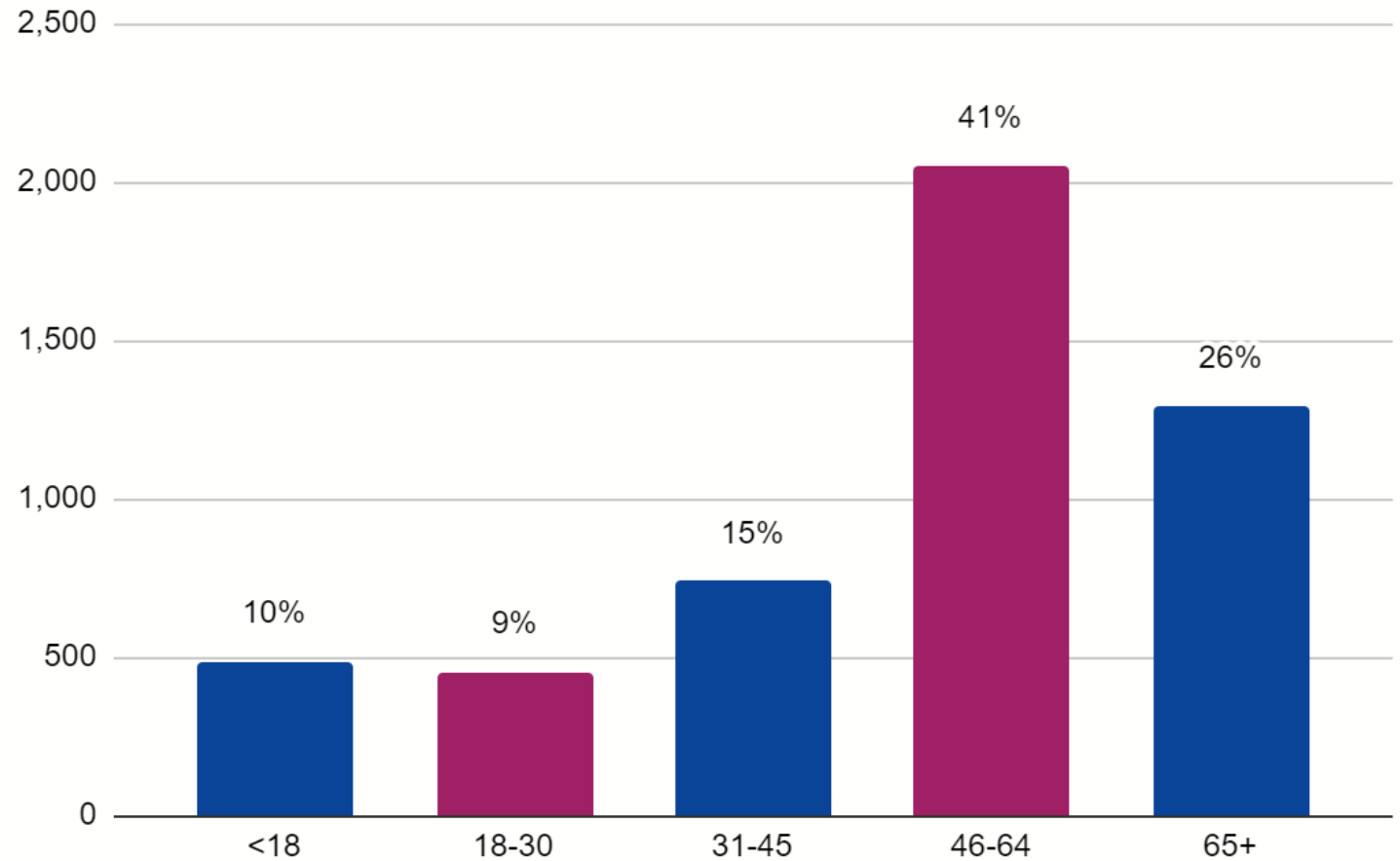
Data collected at baseline and annually:

- Demographics
- FH History, physical findings – arcus, xanthomas, etc.
- Family history
- Vitals (ht., wt., BP)
- Medical History/ASCVD history/COVID-19 History
- Genetic testing results
- Imaging Results
- Laboratory Results
- Current lipid lowering medications/apheresis

Subject Demographics

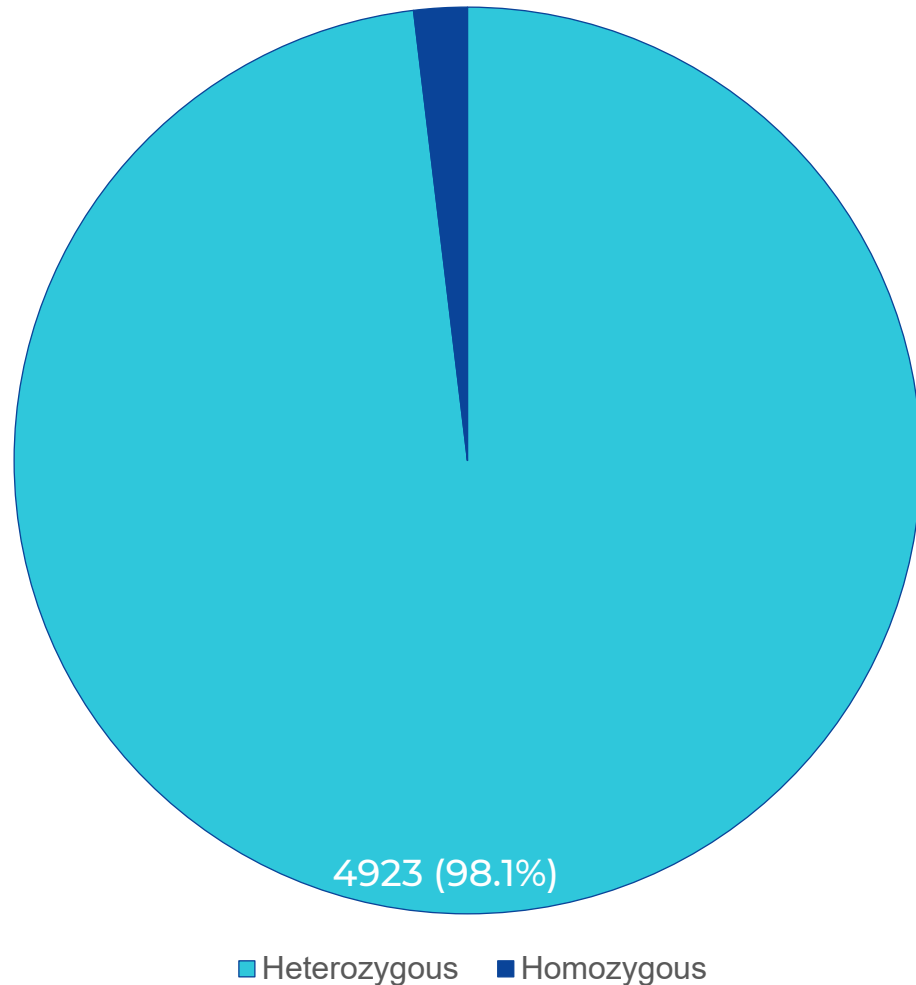
Age (n = 5019)	
Median Age	55
% of Individuals <18	10% (487)
Gender (n = 5016)	
Male	39% (1972)
Female	61% (3044)
Race (n = 5019)	
White	84% (4224)
Black/African American	6% (312)
Asian	3% (149)
Other	7% (334)
Ethnicity (n = 5019)	
Hispanic	4% (208)

Age Distribution within the CASCADE FH Registry



Diagnosis & Comorbidities

FH Diagnosis



Adult Comorbidities

Past Medical History	HeFH (n = 4923)	HoFH (n = 96)
History of Smoking	30% (1470)	26% (25)
Smoking at Enrollment	6% (294)	6% (6)
BMI > 30	31% (1538)	34% (33)
Hypertension	38% (1884)	34% (33)
Diabetes	12% (573)	8% (8)
Peripheral Vascular Disease	4% (216)	11% (11)
Aortic Valve Stenosis	3% (150)	22% (21)
Prior CAD	30% (1472)	58% (56)
Prior MI	11% (547)	19% (18)
Prior Stroke	2% (123)	3% (3)
Prior TIA	3% (136)	2% (2)
Prior PCI or Stent	14% (713)	24% (23)
Prior CABG	9% (465)	28% (27)

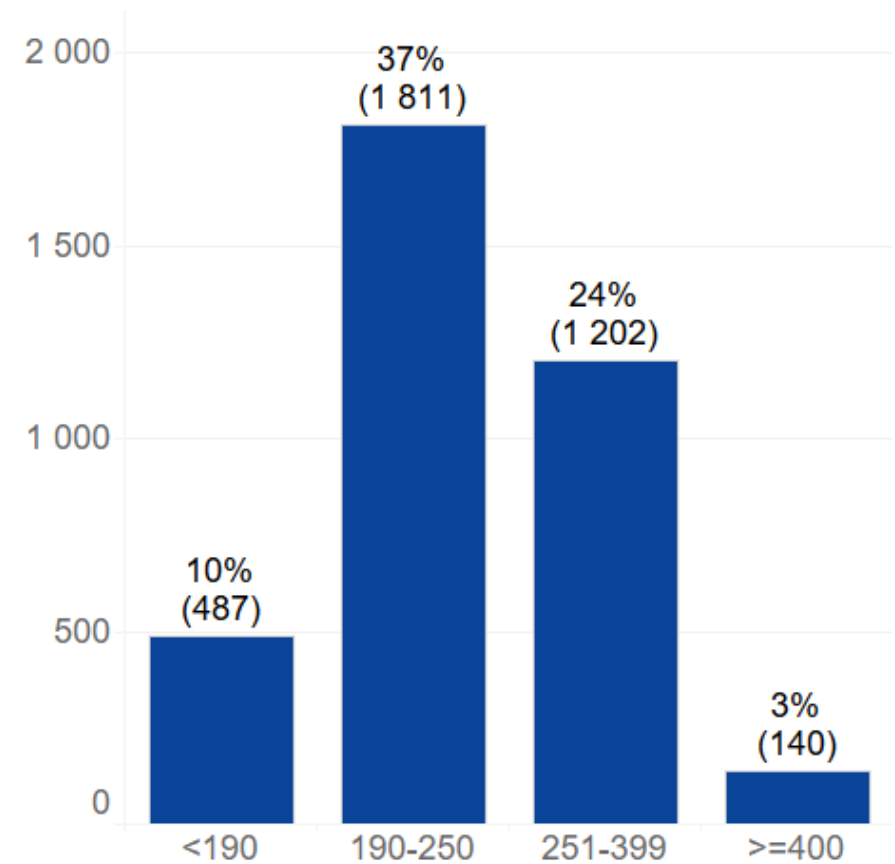
Reported Family History and Diagnoses

Reported Family Hx. and Dx. (n=5019)	
History of FH	32% (1594)
• HeFH	• 72% (1155/1594)
• HoFH	• 3% (44/1594)
• Unknown	• 25% (395/1594)
Hypercholesterolemia	75% (3771)
Premature CVD	52% (2629)
Peripheral Vascular Disease	5% (232)
Xanthoma	6% (303)
Corneal Arcus	3% (141)

HeFH Age, Physical Findings, Genetic Testing, and Procedures (n = 4923)

	% of Individuals (n)
Age <18 at Enrollment	10% (473)
Tendon Xanthomas	21% (1016)
Underwent Genetic Testing	15% (717)
• Positive Genetic Mutation	• 74% (534/717)
Ever Received Lipid Apheresis	2% (90)

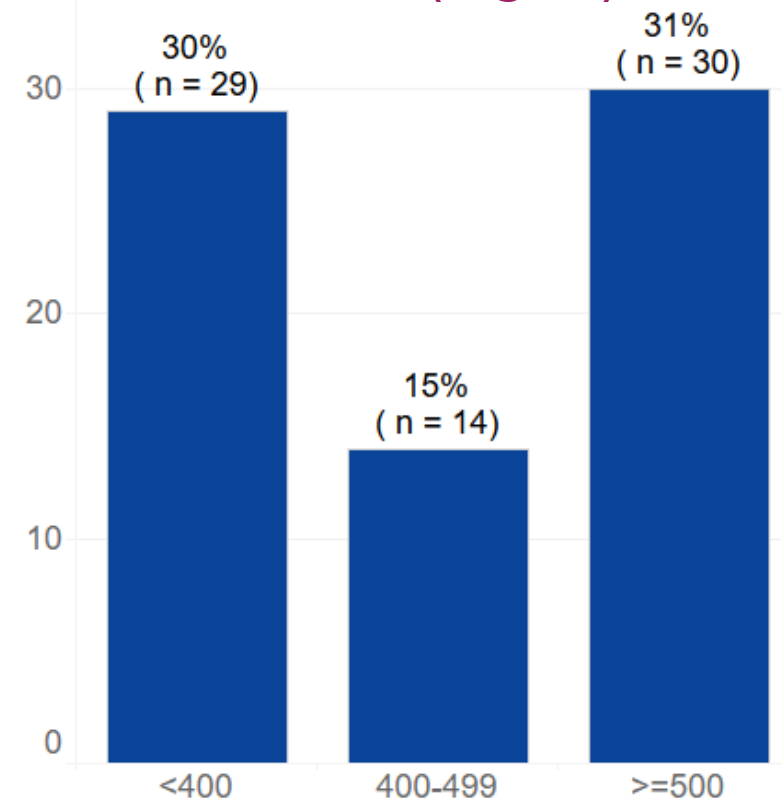
Pre-Treatment LDL-C (mg/dL) Distribution



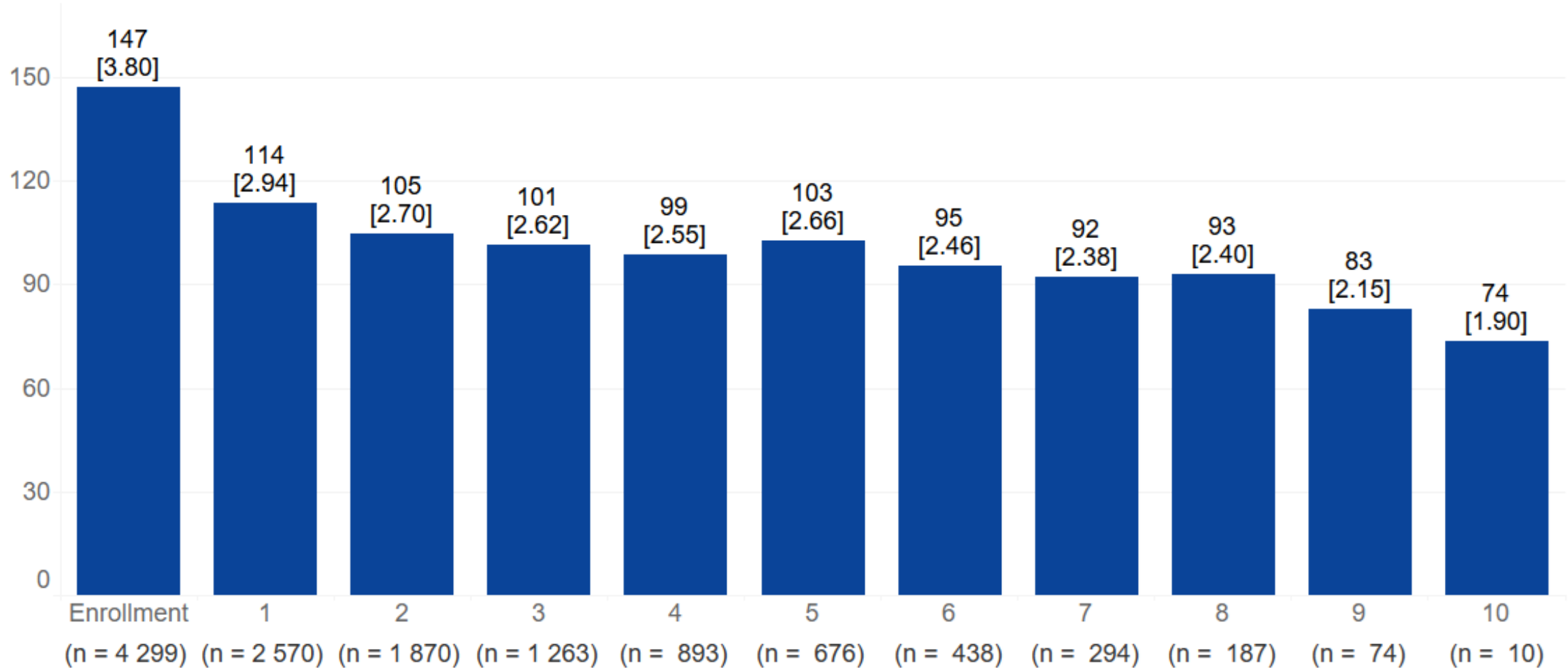
HoFH Age, Physical Findings, Genetic Testing, Medications, and Procedures (n = 96)

	% of Individuals (n)
Age <18 at Enrollment	15% (14)
Tendon Xanthomas	51% (49)
Underwent Genetic Testing	31% (30)
• Positive Genetic Mutation	• 97% (29/30)
Ever Received Lomitapide or Evinacumab	24% (23)
Ever Received Lipid Apheresis	30% (29)

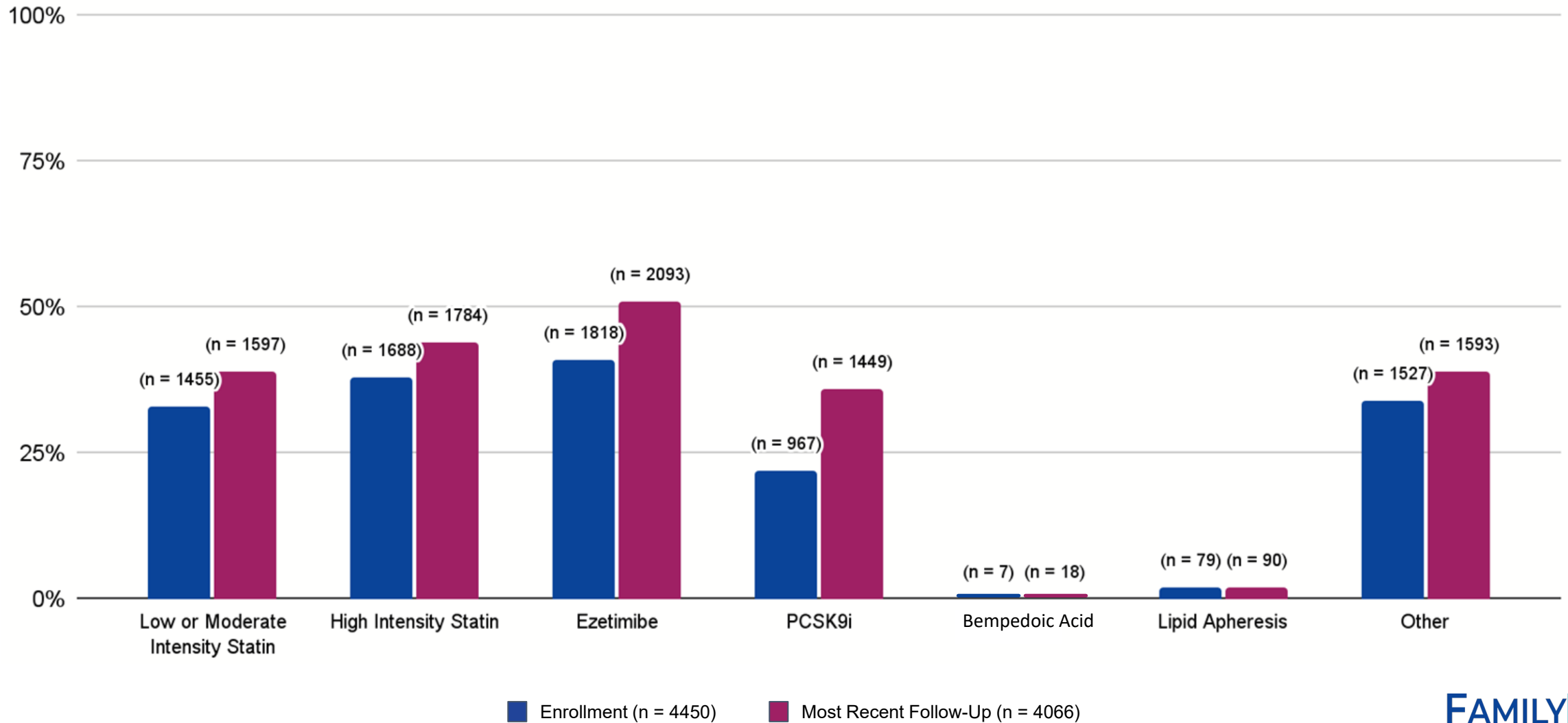
Pre-Treatment LDL-C (mg/dL) Distribution



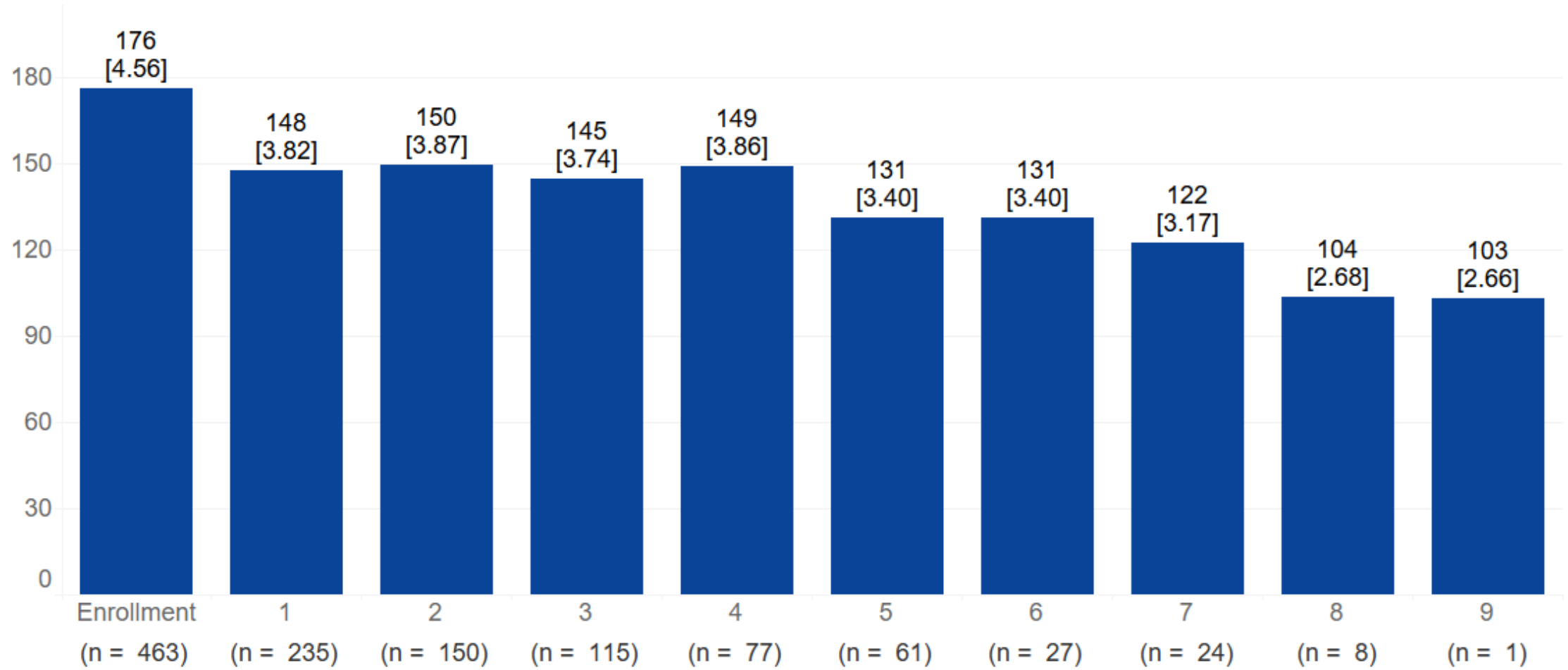
Average LDL-C by entry (mg/dL) [mmol/L] • HeFH Adults



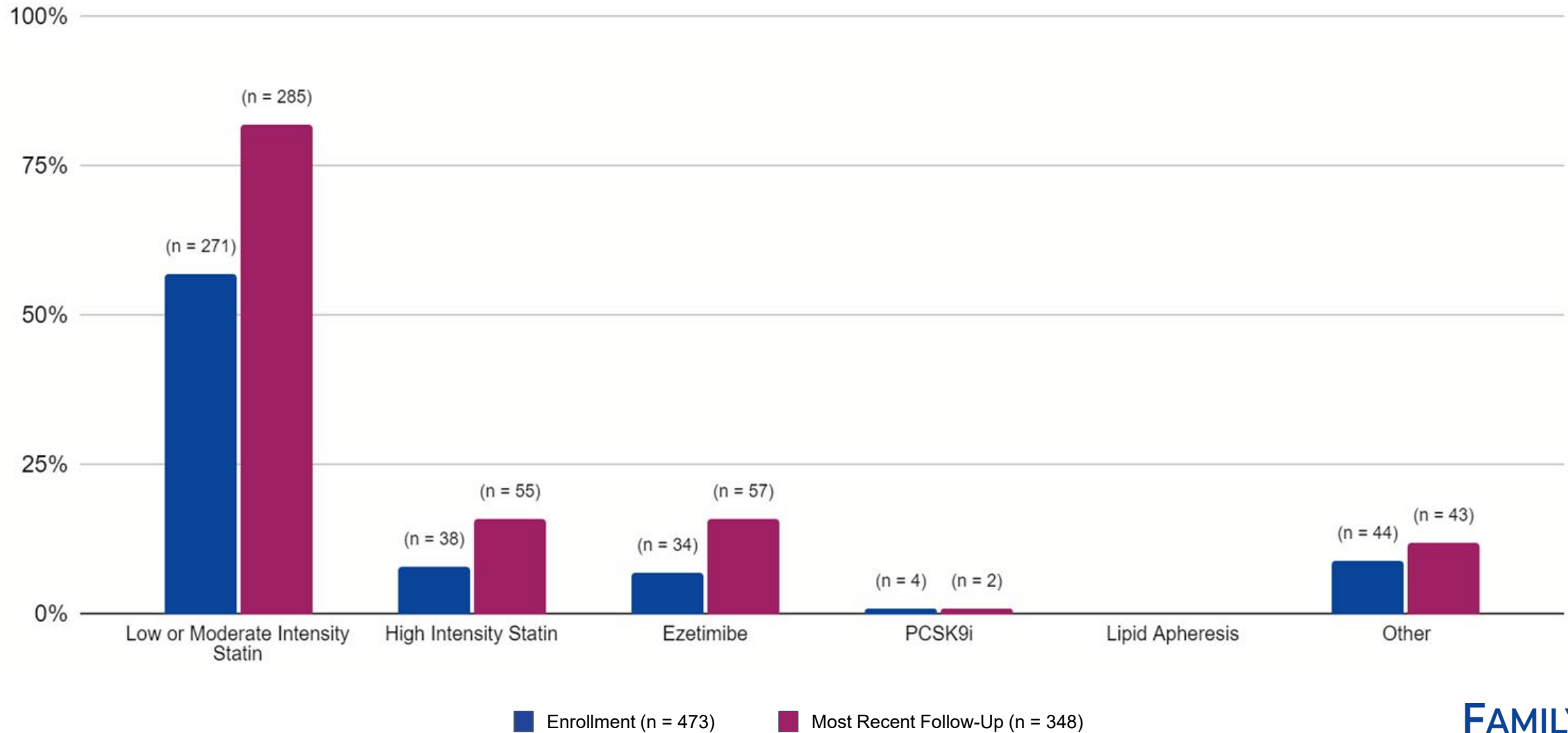
Trends in medication usages • HeFH Adults



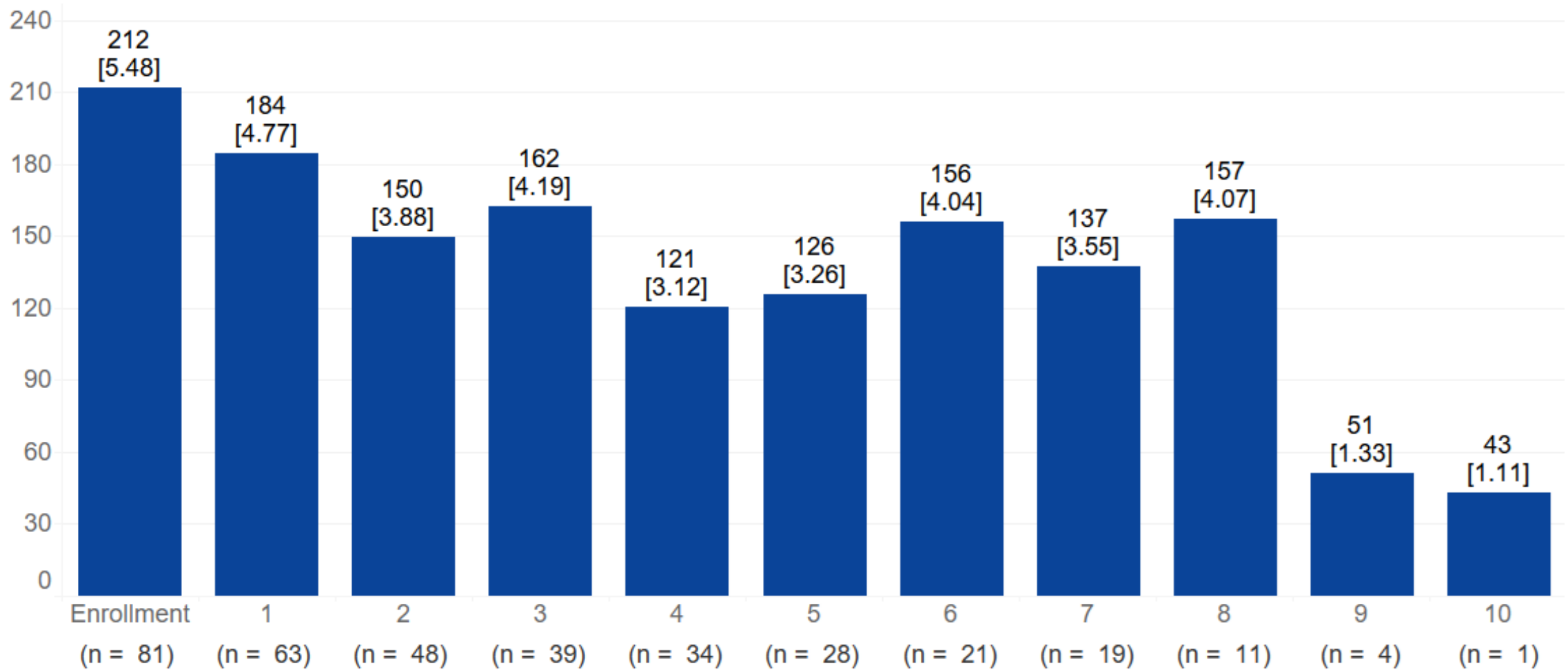
Average LDL-C by entry (mg/dL) [mmol/L] • HeFH Children



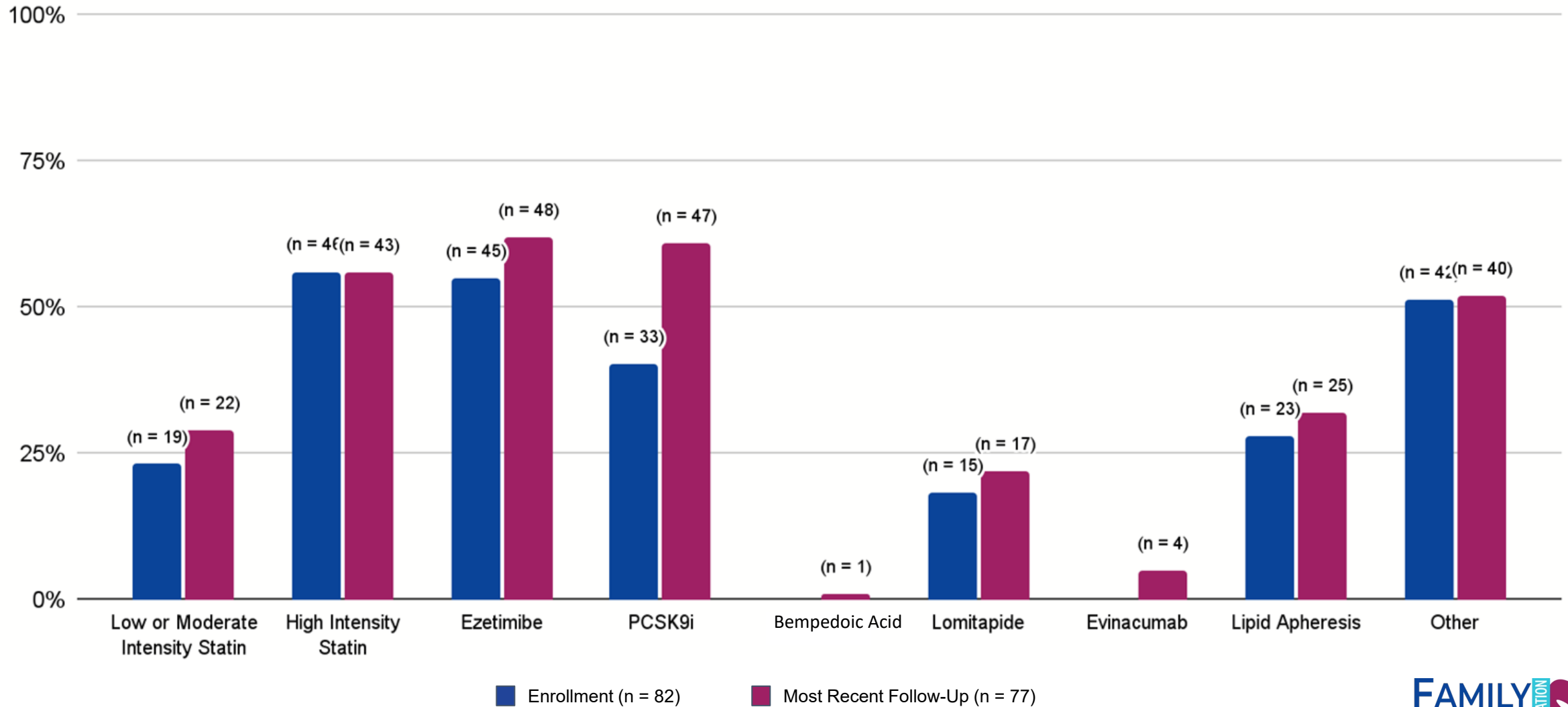
Trends in medication usages • HeFH Children



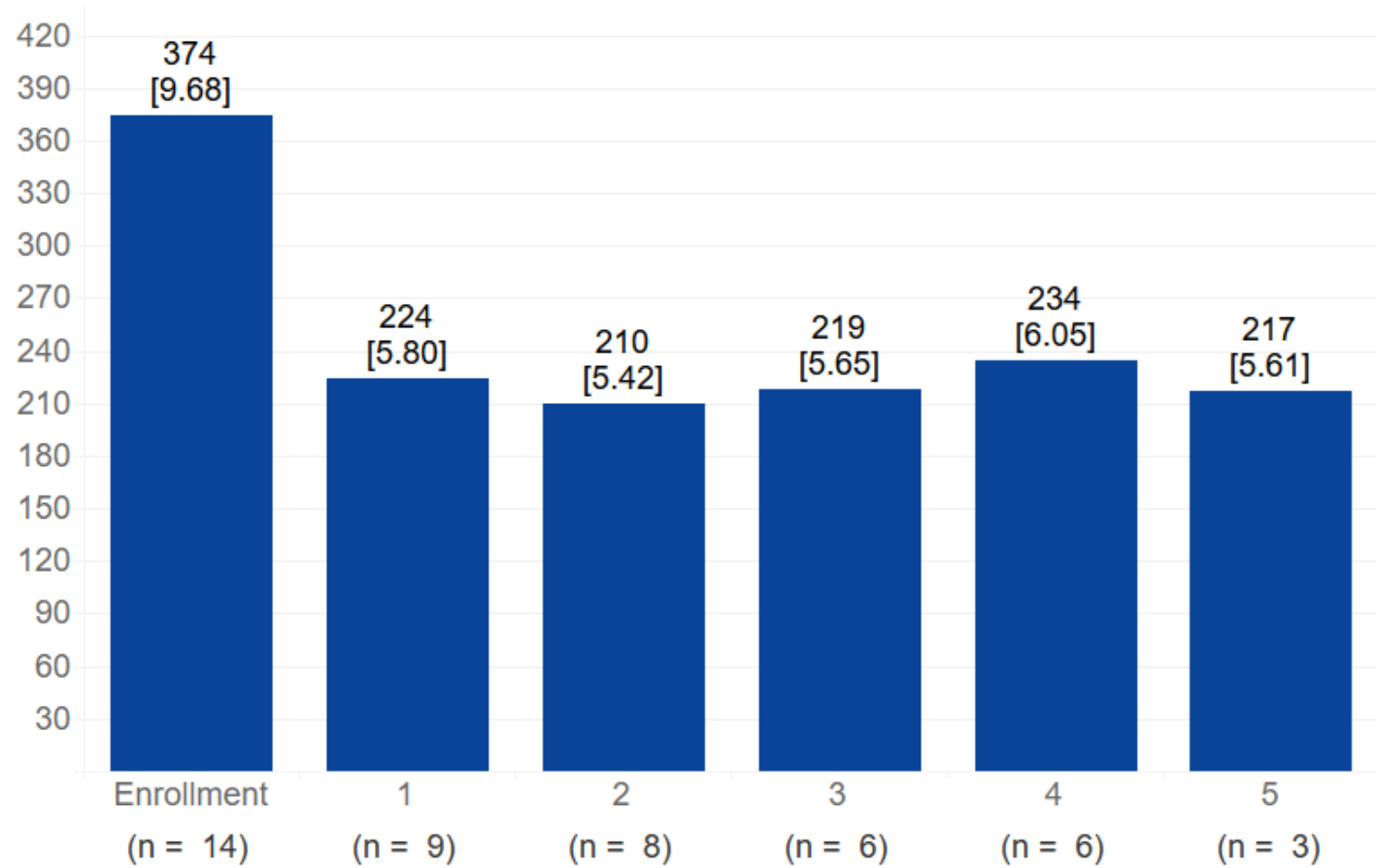
Average LDL-C by entry (mg/dL) [mmol/L] • HoFH Adults



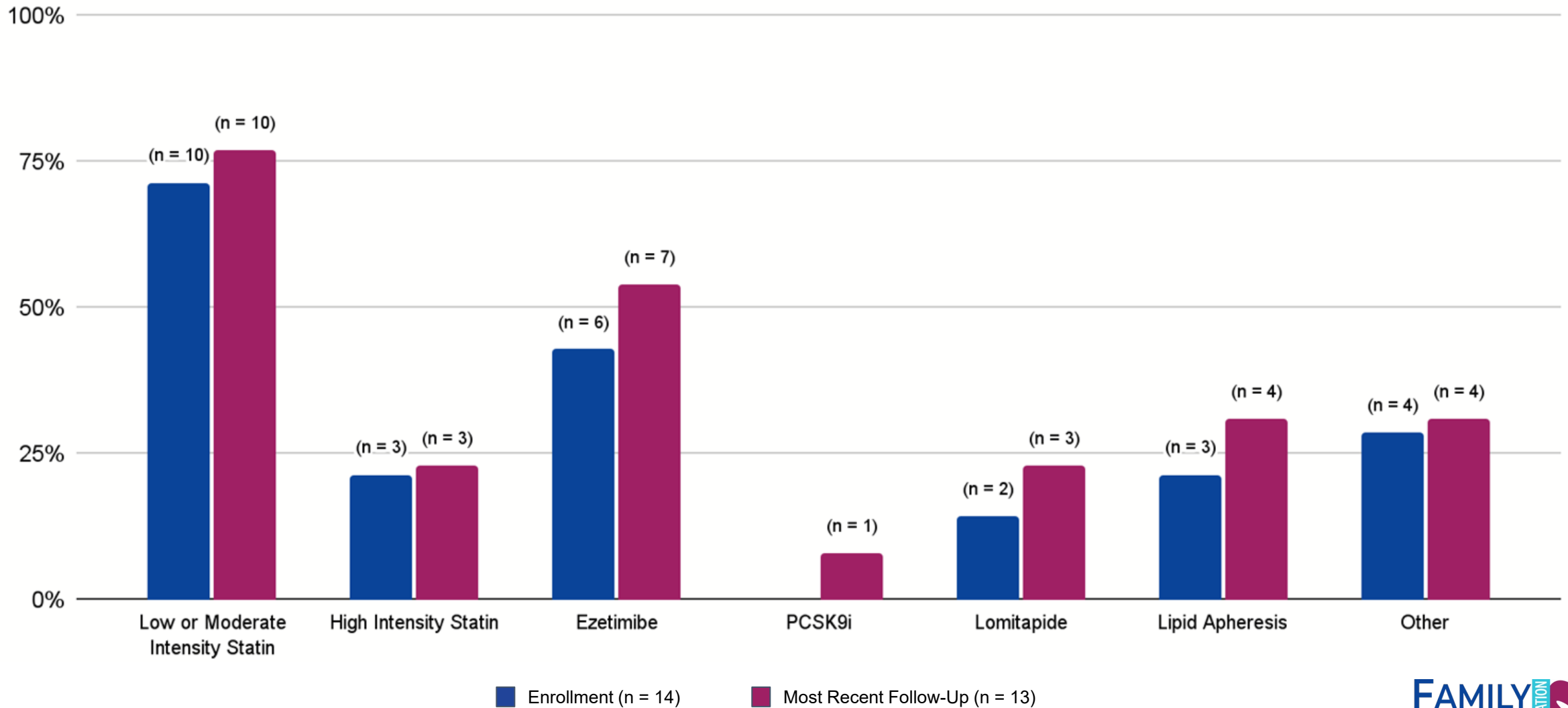
Trends in medication usages • HoFH Adults



Average LDL-C by entry (mg/dL) [mmol/L] • HoFH Children

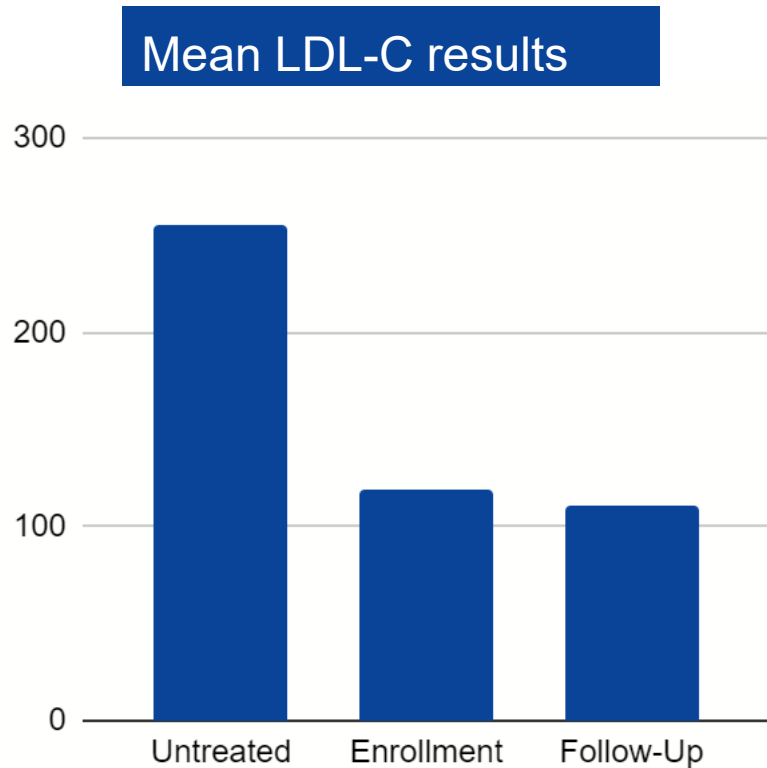


Trends in medication usages • HoFH Children

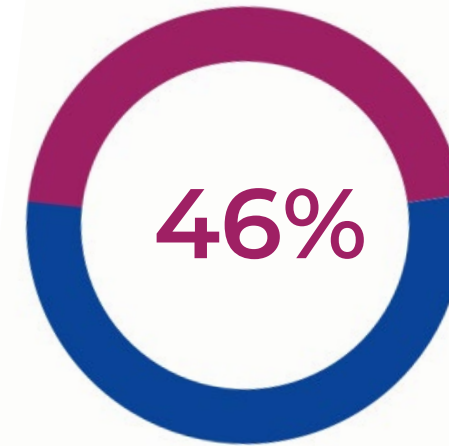


Individuals with HeFH and no ASCVD

Adults under specialty FH care were able to further lower LDL-C, but not far enough



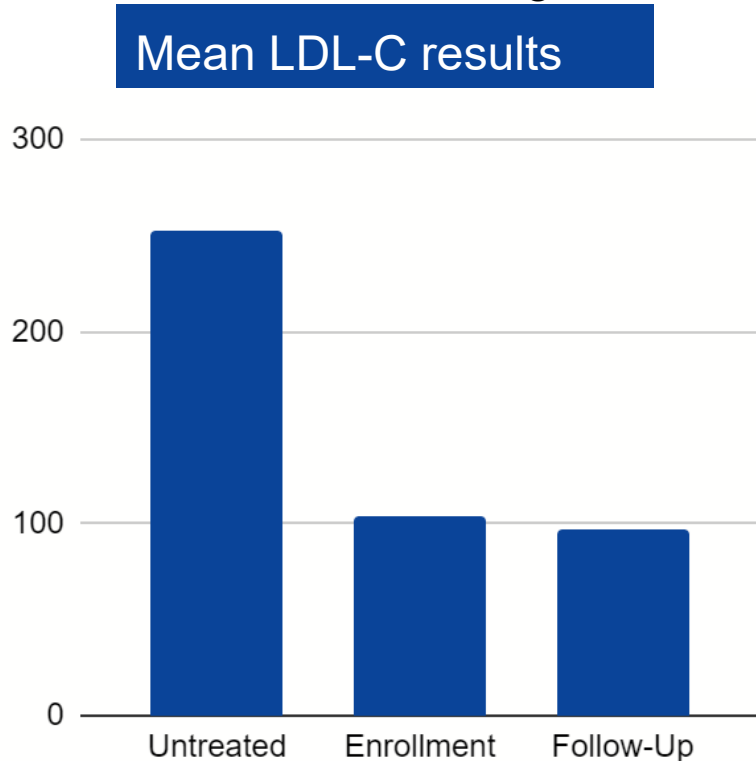
Individuals who had no prior cardiovascular disease



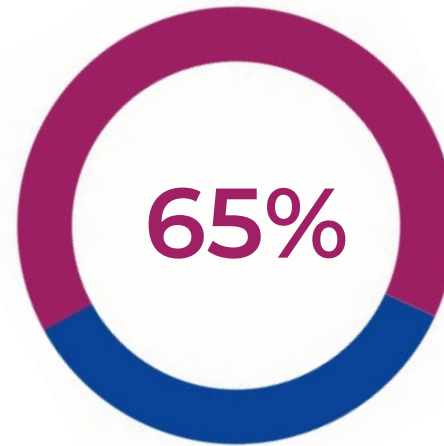
achieved
LDL-C < 100
mg/dL
[2.6 mmol/L]

Individuals with HeFH and ASCVD require 2 or more lipid lowering therapies to reach LDL-cholesterol guidelines

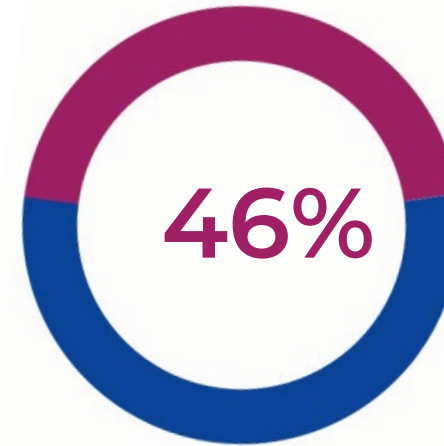
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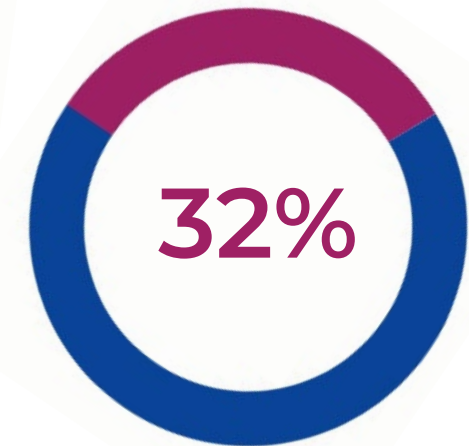
Individuals who had prior cardiovascular disease were more likely to meet targets because they were on 3-6 lipid-lowering therapies including PCSK9 inhibitors or were receiving lipoprotein apheresis



achieved
LDL-C < 100
mg/dL
[2.6 mmol/L]



achieved
LDL-C < 70
mg/dL
[1.8 mmol/L]



achieved
LDL-C < 55
mg/dL
[1.4 mmol/L]












Events reported

	N of Individuals
Covid pts	130
Deaths	57
Pts lost to follow up	502

Events reported

Cardiac events	Primary event on Enrollment	Primary event on Follow-Up Visit	Secondary event on Follow-Up Visit	Unique Subjects	Unique Events
MI	565	22	26	591	621
TIA	138	10	6	151	154
Stroke	126	0	20	144	147
PCI/Stent	736	54	0	757	832
CABG	492	19	0	506	515
Revascularization(Coronary, Peripheral or Carotid Artery)	9	17	0	26	36
Aortic Valve Replacement	21	9	0	30	31

CASCADE FH Publications

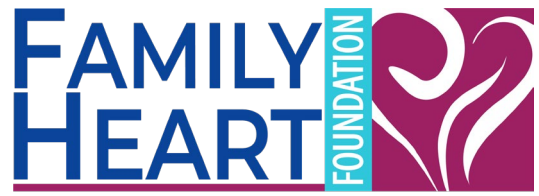
2021		A proof-of-concept study of cascade screening for Familial Hypercholesterolemia in the US, adapted from the Dutch model
2020		Patient acceptance of genetic testing for familial hypercholesterolemia in the CASCADE FH Registry
		Children with Heterozygous Familial Hypercholesterolemia in the United States: Data from the Cascade Screening for Awareness and Detection-FH Registry
2019		Longitudinal low density lipoprotein cholesterol goal achievement and cardiovascular outcomes among adult patients with familial hypercholesterolemia: The CASCADE FH registry
2017		The Role of Registries and Genetic Databases in Familial Hypercholesterolemia
		Health disparities among adult patients with a phenotypic diagnosis of familial hypercholesterolemia in the CASCADE-FH patient registry
2016		US Physician Practices for Diagnosing Familial Hypercholesterolemia: Data from the CASCADE FH Registry
		Treatment Gaps in Adults with Heterozygous Familial Hypercholesterolemia in the United States: Data from the CASCADE FH Registry
2015		Diagnosing Familial Hypercholesterolemia in the United States: Results from the CASCADE FH Patient Registry
		The Rationale and Design of the CASCADE FH Registry: Expert Analysis
2014	24 	Rationale and Design of the Familial Hypercholesterolemia Foundation CAscade SCReening for Awareness and DEtection of Familial Hypercholesterolemia Registry

Faces of FH





Thank you



Lipoprotein(a) & Familial Hypercholesterolemia