

# Familial hypercholesterolemia patients at high risk for cardiovascular events

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## Goal achievement and cardiovascular outcomes among adults with familial hypercholesterolemia: CASCADE FH® Registry

1900 patients with Familial Hypercholesterolemia Median age 56 years Mean age at FH diagnosis 50±18 years

61% female; Untreated LDL-C 249 mg/dL

High rate of cardiovascular disease at enrollment

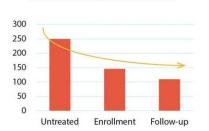
1196 without diagnosed cardiovascular disease

704 with diagnosed cardiovascular disease

Majority of FH individuals did NOT meet guideline-based LDL cholesterol targets despite 2/3 of patients taking two or more lipid-lowering medications

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

#### Mean LDL-C Results Over Time



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



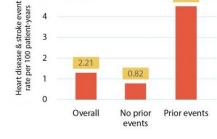
did NOT achieve

LDL-C <100 mg/dL LDL-C <70 mg/dL

#### FH is high-risk and should be considered a cardiovascular disease risk equivalent

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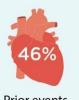
Occurrence of cardiovascular events



## Estimated 10-year cardiovascular event rate is high









No prior events

Prior events



The FH Foundation established and maintains the CAscade SCreening for Awareness and DEtection of Familial Hypercholesterolemia (CASCADE FH®) Registry. The Registry collects comprehensive longitudinal data to better understand outcomes of familial hypercholesterolemia diagnosis and treatment.

Duell, PB et al. Atherosclerosis, Volume 289, 85 - 93

Familial Hypercholesterolemia patients at high-risk for cardiovascular events



despite treatment. Earlier diagnosis and more aggressive cholesterol lowering may reduce the risk according to new data from the FH Foundation's CASCADE FH Registry. Credit: The FH Foundation

Individuals with familial hypercholesterolemia (FH) were able to lower their cholesterol under FH specialty care, but many are still not meeting LDL-cholesterol targets, according to the FH Foundation's CASCADE FH Registry. In new research published in the October issue of *Atherosclerosis*, 52 percent of adults with FH still had LDL-cholesterol over 100 mg/dL despite being on multiple cholesterol-lowering treatments. In addition, these individuals with FH had high heart attack, stroke, and vascular procedure rates, with an annualized cardiovascular event rate of 2.21 per 100 patient years overall, and 4.57 if an individual had a prior cardiovascular event.

"These data from the CASCADE FH Registry demonstrate that individuals with FH are truly a vulnerable, high-risk population for future <a href="heart">heart</a> and stroke events, despite being on multiple treatments," said P. Barton Duell, MD, Professor of Medicine, Director, Lipid-Atherosclerosis Laboratory, Center for Preventive Cardiology, Knight Cardiovascular Institute, Oregon Health and Science University. "These data suggest that FH patients without known heart disease may actually have a risk of heart attack that is comparable to the risk in patients without FH who have already had a heart attack. Most individuals with FH will require multiple medications to adequately lower the LDL-cholesterol level to below goal. Adding additional medications to achieve an LDL-C level that can help prevent a heart attack, particularly for those who already have heart disease, needs to become the standard of care."

While cardiovascular event rates were high overall, rates of major



cardiovascular events were almost six times higher among individuals with prior cardiovascular disease compared to those without. These data underscore the need to treat most patients with FH much more aggressively, possibly with an optimal LDL-C target that may be

"These five years of data from across the United States underscore the impact of life-long exposure to LDL cholesterol in the FH population," said Katherine Wilemon, Founder and CEO of the FH Foundation. "The vast majority of individuals with FH in the United States continue to live without a diagnosis, barring them from the opportunity to receive appropriate care. Finding these individuals and initiating intensive treatment early in life is critical to preventing <a href="heart disease">heart disease</a>."

The results confirmed very late diagnosis of FH, at age 50 (plus or minus 18 years). Lack of awareness of FH in the public and <u>medical</u> <u>community</u> contributes to low diagnosis rates in the United States.

The results also highlighted the broad number of treatments that are now available for FH and the benefit individuals received by being on multiple therapies. The majority of patients are on three to six treatments to lower their LDL-C. Specifically, the use of high intensity statins and PCSK9 inhibitors, as well as treatment with LDL apheresis, at enrollment were associated with a higher likelihood of reaching LDL-C goals. Individuals who achieved more than a 50 percent reduction in LDL-C took more lipid lowering therapies during follow-up, compared to those who achieved less than a 50 percent reduction.

"Today, there are more safe and effective treatments for individuals with FH. With multiple LDL-C lowering drugs, plant sterols, LDL apheresis and exciting experimental therapies in development, individuals should advocate with their healthcare providers to add treatments until their LDL targets are achieved," added Samuel Gidding, MD, chief medical officer for the FH Foundation.



**More information:** P. Barton Duell et al, Longitudinal low density lipoprotein cholesterol goal achievement and cardiovascular outcomes among adult patients with familial hypercholesterolemia: The CASCADE FH registry, *Atherosclerosis* (2019). DOI: 10.1016/j.atherosclerosis.2019.08.007

## Provided by The Familial Hypercholesterolemia Foundation

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