Serving the Underserved: Are We Overlooking HoFH Patients?
Financial Disclosure

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Background

- Familial hypercholesterolemia (FH) is one of the most commonly occurring genetic disorders in the world.
- It is estimated that 1 in 250 Americans have heterozygous FH and about 1 in 160,000 to 300,000 have homozygous FH (HoFH).
- Homozygotes are often unresponsive to dietary and pharmacologic intervention and often develop clinically significant ASCVD before reaching the age of 30.
- Most FH patients remain undiagnosed and untreated.
Background

- The National Lipid Association (NLA) conducted a survey to evaluate the knowledge and practice of primary care and other clinicians for patients with HoFH.
- These clinicians are usually the first in the healthcare community to see these patients.
- The initiative was led by a steering committee comprised of several NLA thought leaders:
Methods

- An independent research and survey vendor was used to conduct the survey.
- The survey was sent out via e-mail to 14,904 medical professionals across the United States (US) from a registry of practitioners who agree to take surveys and who matched the specialty requirement.
- The survey was fielded from June 26, 2018 to July 16, 2018.
- A total of 504 clinicians completed the survey.

Screening Criteria

- US-based based clinicians
- Currently treating patients with elevated LDL-cholesterol
- Licensed to prescribe medication
- Eligible medical disciplines:
  - Physician
  - Nurse Practitioner
  - Physician Assistant
- Eligible specialty groups:
  - Family Medicine
  - General Practice
  - Internal Medicine
  - Cardiology
Survey Participants by Region

Northeast  South  Midwest  West
24%       35%     23%       18%
Respondent Demographics

Over four-fifths of respondents (85%) were physicians and almost all respondents (99%) reported a medical area of practice in primary care and general medicine.
Respondent Demographics

The overwhelming majority of respondents do not consider themselves lipid specialists (88%) nor are they certified by the American Board of Clinical Lipidology (ABCL) or Accreditation Council for Clinical Lipidology (ACCL) (92%).

Do You Consider Yourself a Lipid Specialist?
- Yes: 88% (442)
- No: 12% (62)

Are You Certified by the ABCL or ACCL?
- Yes: 8% (39)
- No: 92% (465)
The majority of respondents (56%) practice in a suburban setting and two-thirds are in private practice.
Very High LDL-C in Respondent Practices

In Your Practice Do You Have:

- Pt(s) with untreated LDL-C of ≥400 mg/dL?
  - Yes: 69%
  - No: 31%

- Pt(s) with treated LDL-C levels >300 mg/dL?
  - Yes: 76%
  - No: 24%

- Family with parent who has LDL-C ≥ 400mg/dL and all offspring have LDL-C 160-400mg/dL?
  - Yes: 59%
  - No: 41%
Diagnosis and Management of Patients with an Untreated LDL-C ≥ 400 mg/dL

53% would diagnose this as HoFH. 70% say their first step would be to treat with medication.
Diagnosis and Management of Patients With a Treated LDL-C >300 mg/dL

The majority no longer recognizes HoFH but does appropriately select adjust medication as next step.

What Would Your Diagnosis Be?

- HoFH: 41%
- HeFH: 32%
- Mixed Hyperlipidemia: 20%
- High Cholesterol due to lifestyle: 5%
- FCS: 2%

What Would Your Next Steps Be?

- Refer to a Specialist: 19%
- Lifestyle Changes and Repeat Lipids in 6 Months: 13%
- Adjust Medication: 68%
- 3rd Choice: 55%
Family with parent who has LDL-C \( \geq 400 \text{mg/dL} \) and all offspring have LDL-C 160-400mg/dL

Diagnosis of HoFH was not improved by knowledge of elevated cholesterol levels in all offspring.
HoFH Patient Demographics

Three-quarters of respondents have a patient that was diagnosed with HoFH, of those, 64% have a patient on a PCSK9 inhibitor.
HoFH Patient Demographics

Of the three-quarters of respondents who report having a patient diagnosed with HoFH, one-quarter (24%) have an HoFH patient on LDL-apheresis.
HoFH Patient Demographics

43% of respondents have a treatment goal of LDL-C <100 mg/dL for those HoFH patients who are free of Clinical ASCVD.
The majority of respondents report diagnosing a patient with HoFH. Of those, almost two-thirds (63%) used clinical criteria to diagnose.
HoFH Patient Risk Assessment

Respondents use *family history* and *LDL-C* most often as the risk factors to determine CVD risk.

What Risk Factors Would You Use to Determine CVD Risk?

- **Family History**: 90%
- **LDL-C**: 87%
- **CAC Scoring**: 66%
- **Lipoprotein(a)**: 55%
- **Non-HDL**: 53%

(Number of Respondents)
HoFH Patient Risk Assessment

Over four-fifths (82%) of respondents used risk calculators to determine if patients were at high risk of ASCVD.

Would You Use Risk Calculators for Determining High Risk of ASCVD?

- Yes: 82% (411)
- No: 18% (93)
Treatment of HoFH Patients

Four-Fifths of respondents would use high dose statins to treat HoFH patients. High dose statins were also ranked the highest of all the therapies (1.5 average).
Access to Lipid Specialists

Two-thirds (63%) of respondents report their practice has access to a lipid specialist.
Access to Lipid Specialists by Respondents Who Do Not Consider Themselves Lipid Specialists

Almost two-thirds of respondents (61%) who do not consider themselves lipid specialists report their practice does have access to a lipid specialist.
Access to LDL-Apheresis Centers

Less than one-third (29%) have access to an LDL-apheresis center.
Patients on LDL-Apheresis by Practice Access to an LDL-Apheresis Center

Access to an LDL-Apheresis Center has a major impact on whether or not a respondent has an HoFH patient on LDL-Apheresis.

Does Your Practice Have Access to an LDL-Apheresis Center?
- Yes: 84% (68)
- No: 16% (13)

Do you have an HoFH patient on LDL-apheresis?
- No: 76% (257)
- Yes: 24% (81)

Does Your Practice Have Access to an LDL-Apheresis Center?
- No: 77% (199)
- Yes: 23% (58)
Access to an LDL-Apheresis Center by Region

Access to an LDL-Apheresis Center is lacking across all regions, with approximately two-thirds of respondents not having access in each of the four US Census Regions.
Access to an LDL-Apheresis Center by Practice Location

Access to an LDL-Apheresis Center is lacking across all locations, with rural practices having the least access.

**Rural**
Does Your Practice Have Access to an LDL-Apheresis Center?
- Yes: 13% (9)
- No: 87% (61)

**Suburban**
Does Your Practice Have Access to an LDL-Apheresis Center?
- Yes: 30% (84)
- No: 70% (197)

**Urban**
Does Your Practice Have Access to an LDL-Apheresis Center?
- Yes: 36% (55)
- No: 64% (98)
Age at Which Respondents Would Start HoFH Patients on LCL-C Lowering Medications

The majority of respondents would start an HoFH patient on LDL-C lowering medications at 19 to 29 years of age, for both males and females (54% and 51%, respectively).

**What Age Would You Start a male Patient on LDL-C Lowering Medication?**

<table>
<thead>
<tr>
<th>Age Group</th>
<th>Number of Respondents</th>
</tr>
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<tbody>
<tr>
<td>&lt;18 Yrs</td>
<td>24%</td>
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<tr>
<td>19-29 Yrs</td>
<td>54%</td>
</tr>
<tr>
<td>30-39 Yrs</td>
<td>17%</td>
</tr>
<tr>
<td>&gt;40 Yrs</td>
<td>5%</td>
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**What Age Would You Start a female Patient on LDL-C Lowering Medication?**

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</tr>
<tr>
<td>19-29 Yrs</td>
<td>51%</td>
</tr>
<tr>
<td>30-39 Yrs</td>
<td>19%</td>
</tr>
<tr>
<td>&gt;40 Yrs</td>
<td>10%</td>
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Age at Which Respondents Would Start HoFH Patients on LCL-C Lowering Medications by Specialty

The age at which respondents would start a patient with HoFH on LDL-C lowering medications remains consistent regardless of the respondent’s medical specialty.

What Age Would You Start a **male** Patient on LDL-C Lowering Medication?

<table>
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<tr>
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<td>28%</td>
<td>26%</td>
</tr>
<tr>
<td>30-39 Yrs</td>
<td>9%</td>
<td>8%</td>
</tr>
<tr>
<td>&gt;40 Yrs</td>
<td>2%</td>
<td>3%</td>
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What Age Would You Start a **female** Patient on LDL-C Lowering Medication?

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<td>4%</td>
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Conclusion

Many clinicians:

- Do not recognize HoFH.
- Do not adequately treat HoFH.
- Do not have access to a lipid specialist.
- Do not have access to a LDL-apheresis center.

There is a need for:

- More education for clinicians in recognizing and treating HoFH
- Greater access to lipid specialists.
- Greater access to LDL-apheresis centers.
Quick EHR Search Terms

- Untreated LDL-C ≥ 400 mg/dL
- Treated LDL-C >300 mg/dL
- Family history of high cholesterol
- Family history of premature CV disease
References

