WEBCAST

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Topline Pivotal Study Results in Spinocerebellar Ataxia

September 23, 2024

Amy, living with SCA
Clinical trial participant continuing open label treatment

Forward-Looking Statements

This presentation includes forward-looking statements within the meaning of the Private Securities Litigation Reform Act of 1995, including statements about Biohaven Ltd. (the "Company") and our planned and ongoing clinical trials, the timing of and the availability of data from those trials, the timing and our decisions to proceed with our planned regulatory filings (including our plans to submit a NDA to the FDA for troriluzole in the treatment of all SCA genotypes in 4Q 2024), the timing of and our ability to obtain regulatory approvals for our product candidates (including the timing of the regulatory approval for troriluzole in order to commercialize SCA in the United States in 2025), the clinical potential utility of our product candidates, alone and as compared to other existing potential treatment options, and the potential advancement of our early phase programs. The use of certain words, including "continue", "plan", "will", "believe", "may", "expect", "anticipate" and similar expressions, is intended to identify forward-looking statements. Investors are cautioned that any forward-looking statements, including statements regarding the future development, timing and potential marketing approval and commercialization of our development candidates, are not guarantees of future performance or results and involve substantial risks and uncertainties. Actual results, developments and events may differ materially from those in the forward-looking statements as a result of various factors including: the expected timing, commencement and outcomes of Biohaven's planned and ongoing clinical trials; the timing of planned interactions and filings with the FDA; the timing and outcome of expected regulatory filings, including the timing and outcome of the NDA for troriluzole; complying with applicable U.S. regulatory requirements; the potential commercialization of Biohaven's product candidates, including the commercialization of SCA in the United States in 2025; and the effectiveness and safety of Biohaven's product candidates. You should, therefore, not rely on these forward-looking statements as representing our views as of any date subsequent to the date of this presentation. Additional important factors to be considered in connection with forward-looking statements are described in the Company's filings with the Securities and Exchange Commission, including within the sections titled "Risk Factors" and "Management's Discussion and Analysis of Financial Condition and Results of Operations". The forward-looking statements are made as of the date of this presentation, and Biohaven does not undertake any obligation to update any forward-looking statements, whether as a result of new information, future events or otherwise, except as required by law. This presentation also contains market data and other information based on industry publications, reports by market research firms or published independent sources. Some market data and information is also based on the Company's good faith estimates, which are derived from management's knowledge of its industry and such independent sources referred to above.

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AGENDA

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Introduction
Matthew Buten
Chief Financial Officer, Biohaven

Opening Remarks
Vlad Coric, M.D.
Chief Executive Officer and Chairman, Biohaven

Spinocerebellar Ataxia Overview
Irfan Qureshi, M.D.
Chief Medical Officer, Biohaven

BHV4157-206-RWE Pivotal Study Results
Melissa Beiner, M.D.
SCA Clinical Lead and Senior Medical Director, Biohaven

Expert Perspective on Topline Results
Jeremy Schmahmann, M.D.
Professor of Neurology, Harvard Medical School
Director, Mass General Hospital Ataxia Center
Medical Research Advisory Board, National Ataxia Foundation

SCA Commercial Planning
John Tilton
Chief Commercial Officer for Rare Disease, Biohaven

Q&A

Opening Remarks

Vlad Coric, M.D.

Chief Executive Officer and Chairman, Biohaven

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Troriluzole 200 mg QD dosed orally in patients with SCA

MET THE STUDY'S PRIMARY ENDPOINT

on the change from baseline on the f-SARA at 3 years in all study population genotypes

Sustained and clinically meaningful treatment benefit out to 3 years across analyses utilizing 2 large independent natural history external controls

- Troriluzole achieved statistically significant superiority on a total of **9 consecutive, prespecified primary and secondary endpoints**
- SCA patients treated with troriluzole showed a **50–70% slowing of disease progression**, representing 1.5–2.2 years delay in disease progression over the 3-year study period

Large safety database demonstrates troriluzole is well tolerated in SCA

New Drug Application submission planned in 4Q 2024

Spinocerebellar Ataxia Overview

Irfan Qureshi, M.D.
Chief Medical Officer, Biohaven

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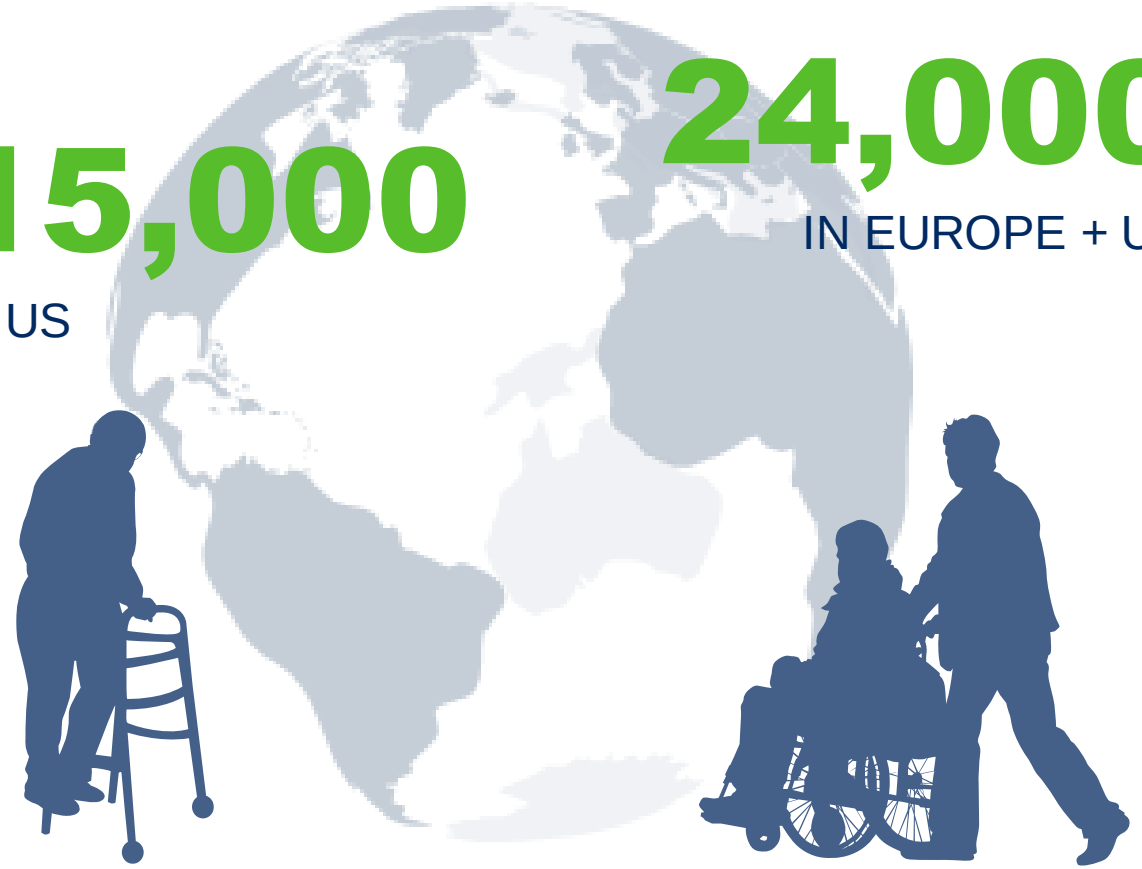
SCA: Rare Progressively Debilitating and Fatal Neurodegenerative Disorder with No Approved Treatment

- Autosomal dominant, progressive, neurodegenerative disease with multiple genotypes[1–3]
- Onset in early adulthood with symptoms leading to severe disability and premature death[3]
- High unmet need and no approved therapies[1,2]

SCA Prevalence[4]

15,000
IN US

24,000
IN EUROPE + UK



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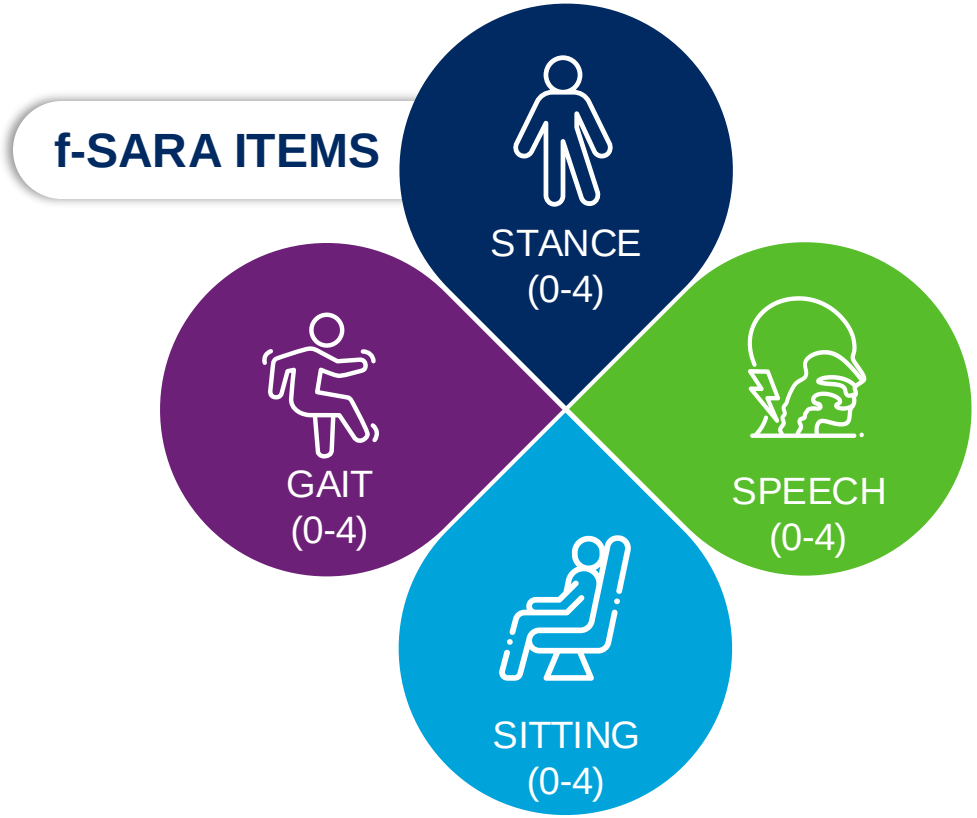
f-SARA: Neurologist-Assessed Scale that Tracks SCA Disease Progression

- Measures 4 core functional items that are clinically meaningful and reflect hallmark symptoms of SCA[5]
- Individual items rated 0–4 with total score range 0–16
- Generally increases (worsens) 0.5 points annually
- Developed with FDA input
- Psychometric and qualitative validation performed according to FDA guidance[5,6]



KEY POINT

f-SARA is an approvable endpoint in SCA

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Troriluzole: Novel Rationally-Designed Therapy for SCA

Strong IP Protection

- Issued NCE Composition of Matter patent expiration anticipated 2041 with extensions

Potential Therapeutic Effects of Troriluzole

Glutamate dysregulation linked to SCA

- Increases intracellular calcium causing excitotoxicity
- Disrupts Purkinje neuron physiology and cerebellar network function
- Leads to neuronal cell death and progressive spinocerebellar degeneration

Troriluzole restores glutamate homeostasis

- Increases glial glutamate uptake and blocks presynaptic release of glutamate[7,8]
- Promotes healthy cerebellar Purkinje neuron functioning[1]
- Reduces excitotoxicity, neuronal damage and cell death

Regulatory Designations for SCA Development

Fast Track in US & Orphan in US/Europe

BHV4157-206-RWE Pivotal Study Results

Melissa Beiner, M.D.
SCA Clinical Lead and Senior Medical Director, Biohaven

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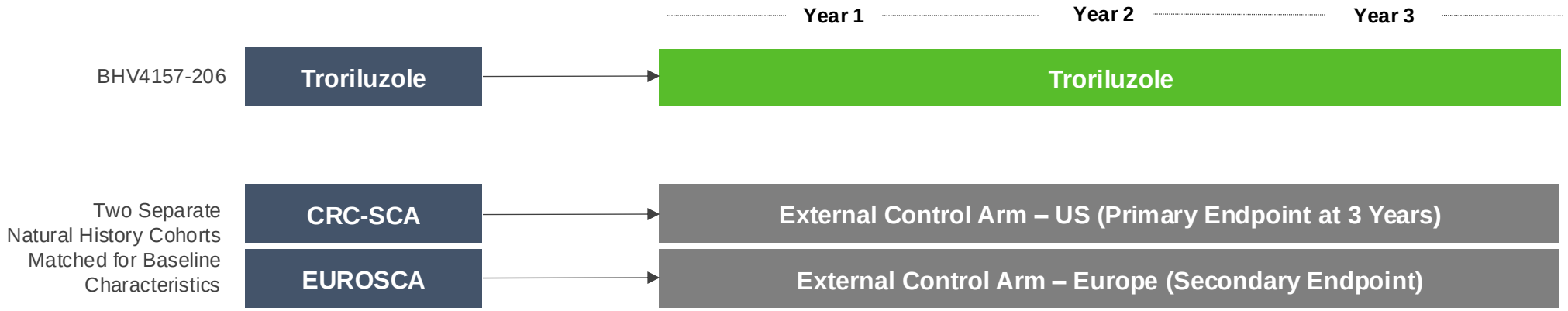
Study BHV4157-206-RWE (NCT06529146): Prespecified Propensity Score Matching with External Control Arm



DESIGN	Propensity Score Matching (up to 3 untreated external control subjects matched to each troriluzole-treated subject)
PRIMARY ENDPOINT	**Total f-SARA Scale** Change from baseline at 3 years in troriluzole-treated subjects vs untreated subjects from US Natural History control (CRC-SCA)
SECONDARY ENDPOINTS INCLUDE	• f-SARA change from baseline at 1 and 2 years vs US Natural History external control (CRC-SCA) • f-SARA change from baseline at 1, 2, and 3 years vs EU Natural History external control (EUROSCA) • f-SARA change from baseline at 1, 2, and 3 years vs pooled US & EU Natural History external control (CRC-SCA & EUROSCA)

Demographic and Baseline Characteristics

	BHV4157-206	CRC-SCA	EUROSCA
n	105	446	358
Age (years), n	105	434	358
mean (SD)	47.6 (13.1)	51.6 (13.8)	47.3 (12.7)
median (range)	49.0 (18, 73)	52.0 (0, 89)	47 (18, 84)
Sex, n	105	446	358
Male (%)	47 (45)	200 (45)	171 (48)
Female (%)	58 (55)	246 (55)	187 (52)
Age at symptom onset (years)			
mean (SD)	37.7 (12.4)	41.2 (13.9)	36.7 (11.8)
median (range)	38 (10, 71)	41 (0, 76)	37 (7, 76)
Genotype (%)			
SCA1	15 (14)	66 (15)	102 (29)
SCA2	31 (30)	94 (21)	141 (39)
SCA3	41 (39)	153 (34)	115 (32)
SCA6	6 (6)	95 (21)	0
SCA7	5 (5)	5 (1)	0
SCA8	3 (3)	19 (4)	0
SCA10	3 (3)	6 (1)	0
Multiple	1 (1)	3 (1)	0
f-SARA			
mean (SD)	4.95 (1.6)	3.97 (3.5)	5.03 (4.1)
median (range)	4.00 (2,10)	3.00 (0,16)	4.00 (0,16)

Full Analysis Set

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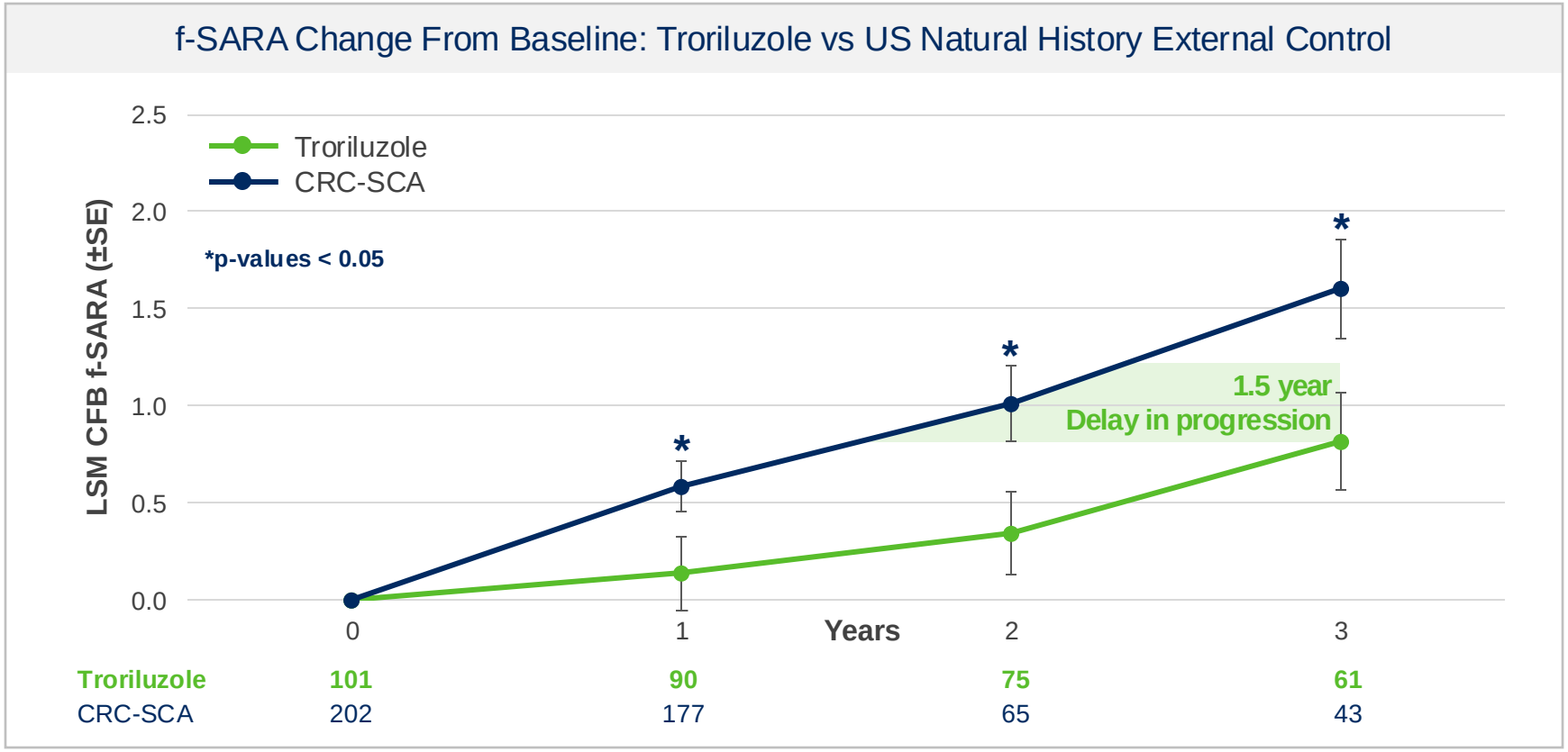
Positive Prespecified Primary and Secondary Endpoints: Troriluzole vs US Natural History External Control



KEY POINT

Troriluzole reduced SCA disease progression by ~50%

CRC-SCA, Clinical Research Consortium for SCA; **EUROSCA,** European registry of SCA; **f-SARA**, Functional Scale for the Assessment and Rating of Ataxia; **LSM,** least squares mean; **PSM**, Propensity Score Matching; **CFB**, Change from baseline

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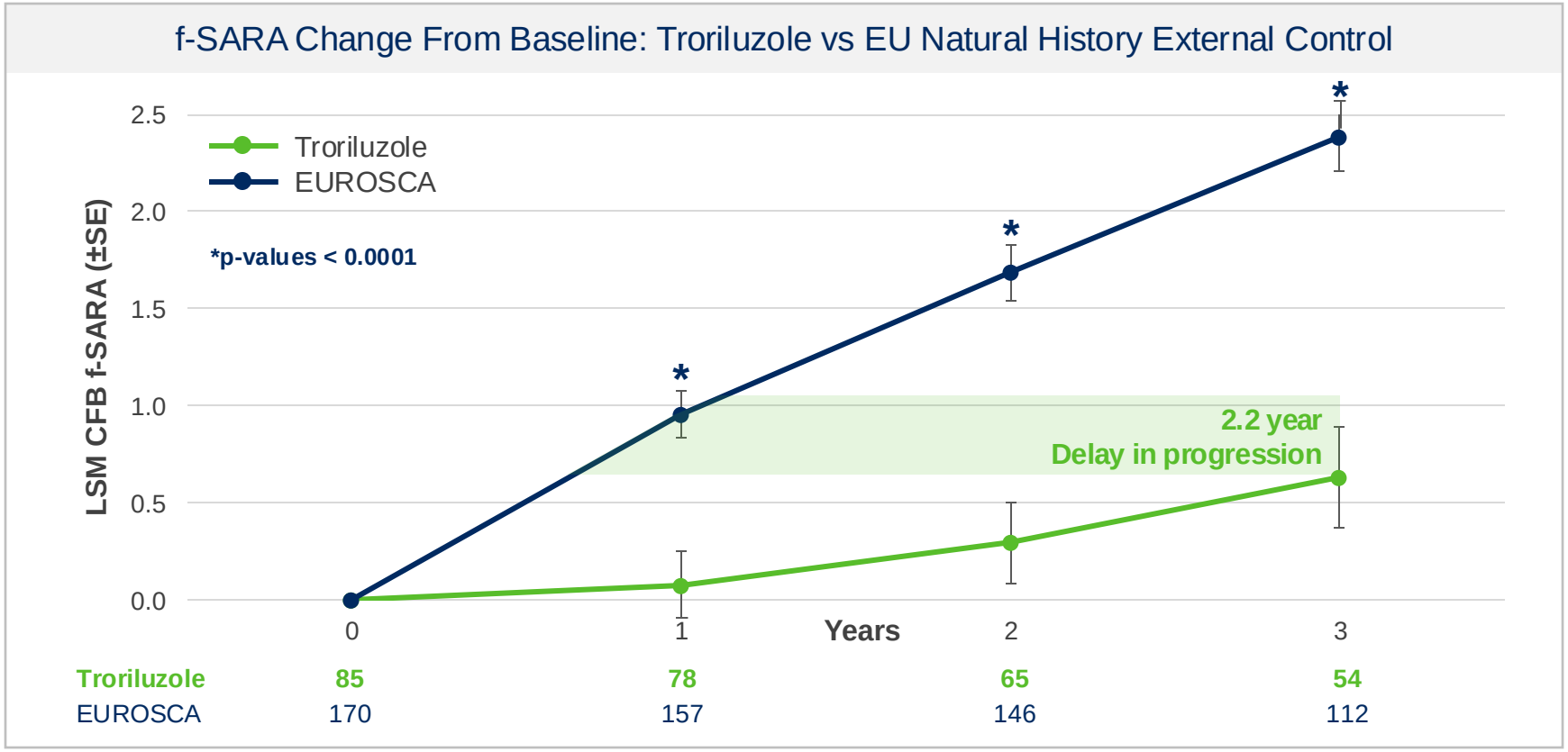
Positive Prespecified Secondary Endpoints: Troriluzole vs Independent EU Natural History External Control



KEY POINT

Troriluzole reduced SCA disease progression by ~70%

CRC-SCA, Clinical Research Consortium for SCA; **EUROSCA,** European registry of SCA; **f-SARA**, Functional Scale for the Assessment and Rating of Ataxia; **LSM,** least squares mean; **PSM**, Propensity Score Matching; **CFB**, Change from baseline

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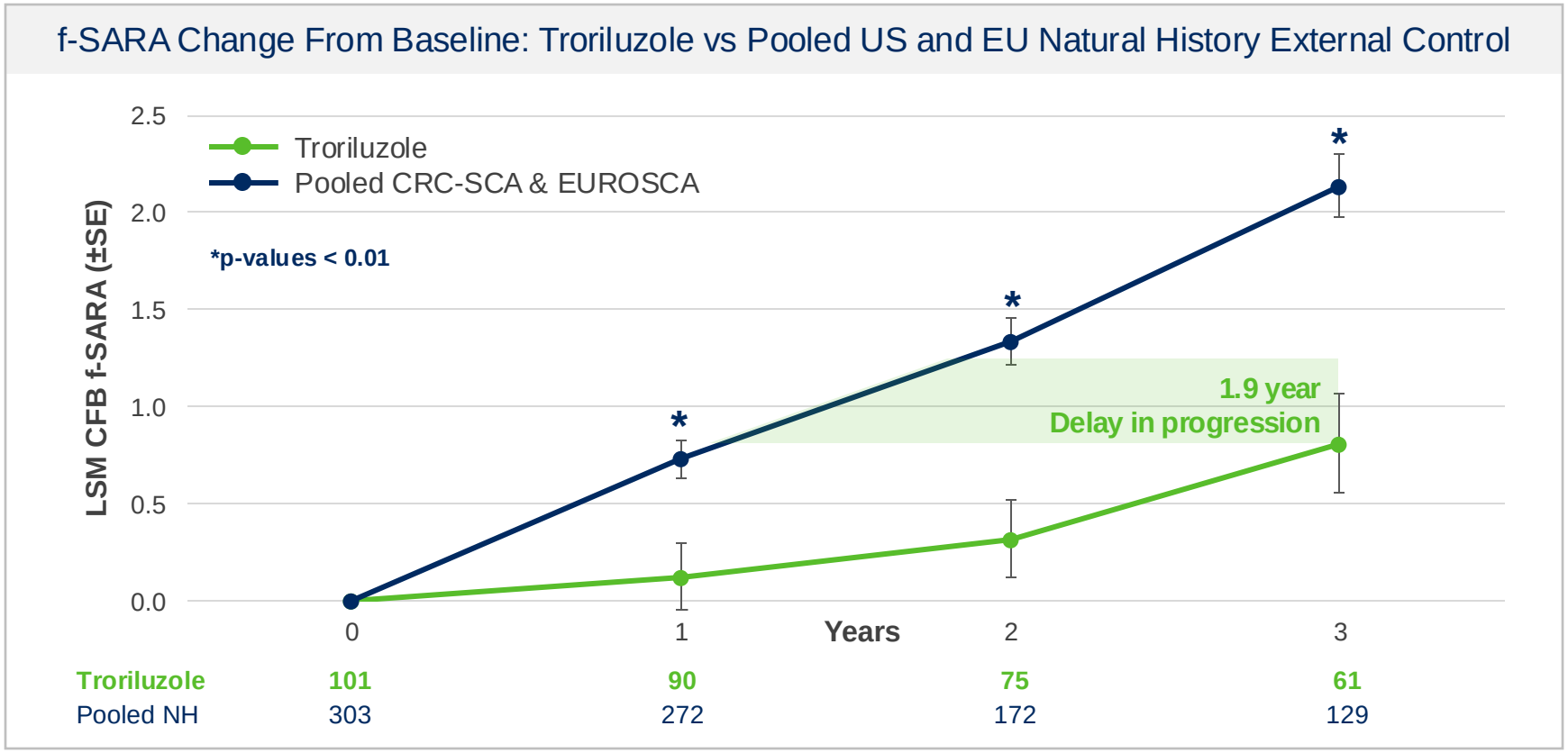
Positive Prespecified Secondary Endpoints: Troriluzole vs Pooled US and EU Natural History External Control



KEY POINT

Troriluzole reduced SCA disease progression by ~60%

CRC-SCA, Clinical Research Consortium for SCA; **EUROSCA,** European registry of SCA; **f-SARA**, Functional Scale for the Assessment and Rating of Ataxia; **LSM,** least squares mean; **PSM**, Propensity Score Matching; **CFB**, Change from baseline

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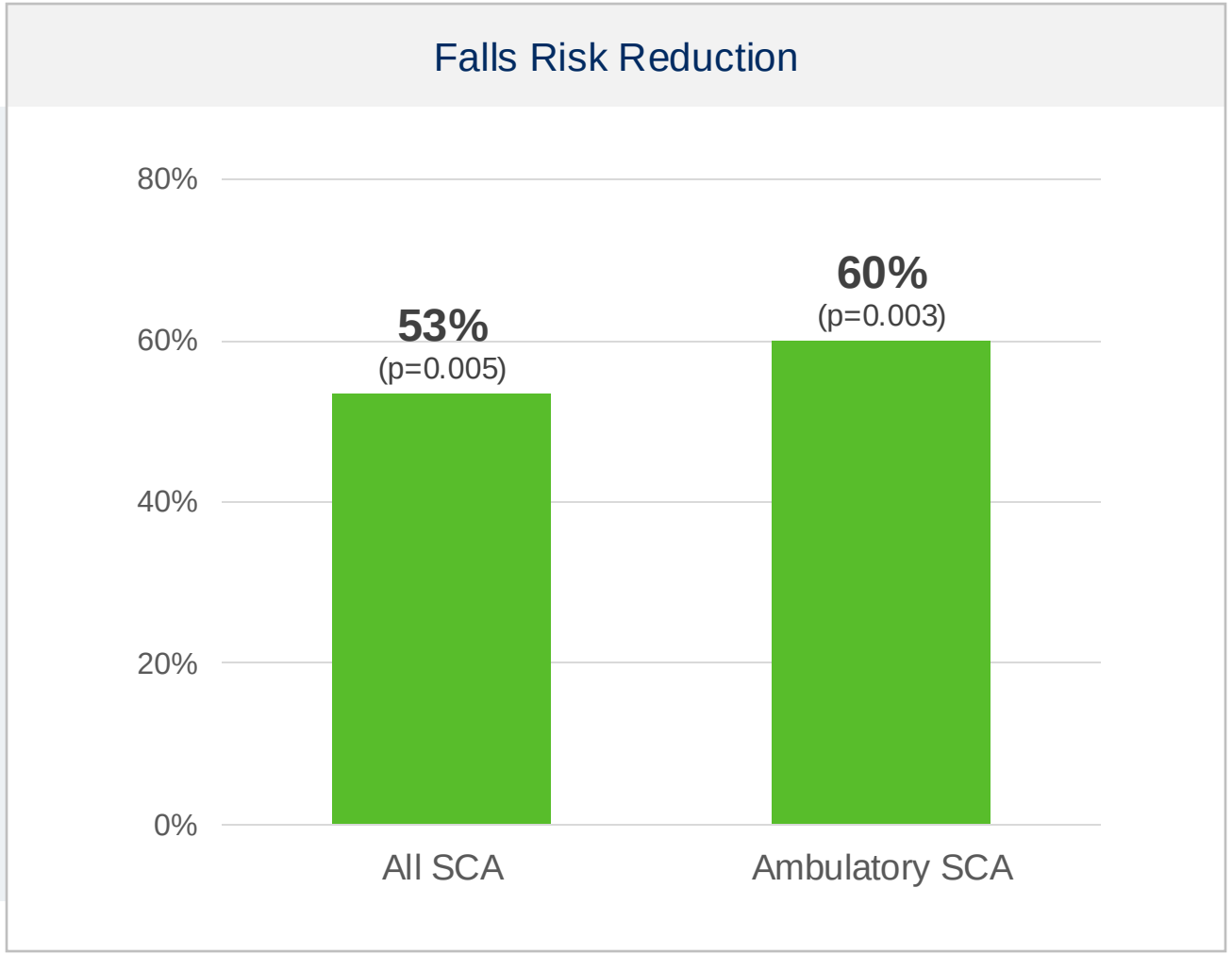
Troriluzole Substantially Reduced Fall Risk in Double-Blind Phase

Burden of Falls in SCA[9–10]

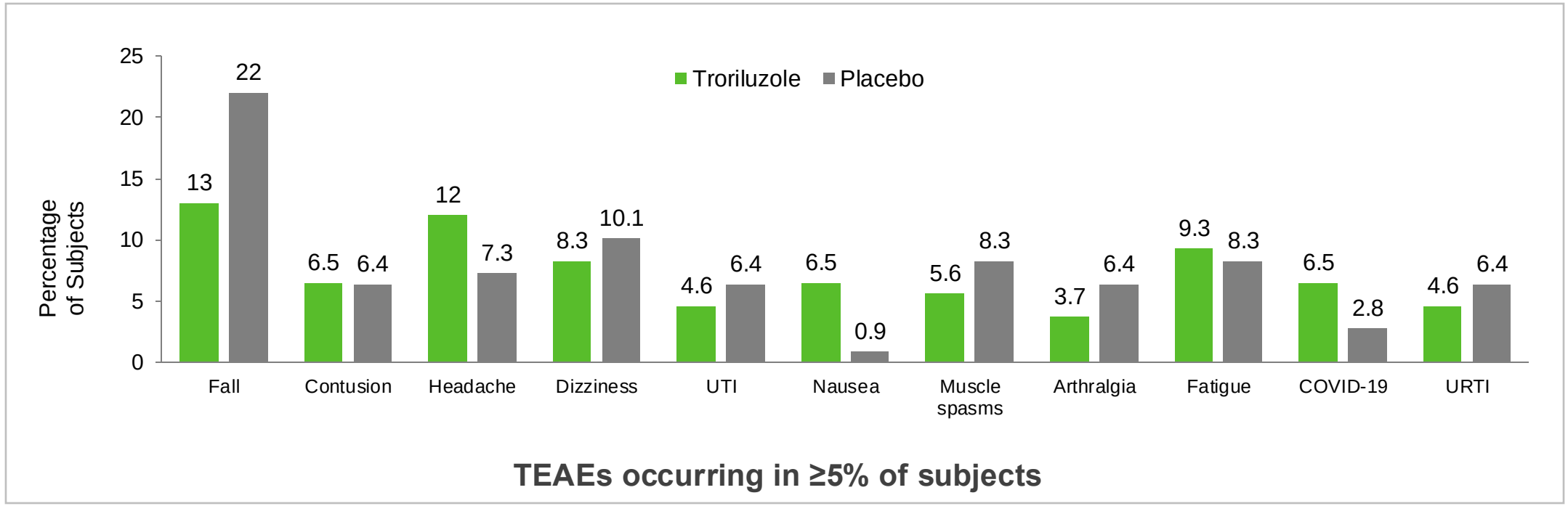
- Most SCA patients (74–84%) report falling in the preceding 12 months
- Falling is associated with a high rate of injury (74%)
- Frequent fallers report more fall-related injuries
- Fall frequency decreases when patients become wheelchair dependent or immobile



* Study BHV4157-206 double-blind phase results; Falls were captured in Study BHV4157-206 as adverse events if reported as "worsening falls" or if the fall resulted in an injury. For the analysis, a generalized linear model was fit using a Poisson family model with a log link function.
** Ambulatory SCA is defined as All SCA subjects who could ambulate without constant assistance (scoring 1 or 2 on the gait item of the f-SARA) at baseline

Troriluzole was Well-Tolerated in Clinical Trials

	Troriluzole N=108	Placebo N=109
Serious TEAE	**6 (5.6)**	8 (7.3)
Severe TEAE	**3 (2.8)**	8 (7.3)
TEAE Leading to Discontinuation	**5 (4.6)**	5 (4.6)



Study BHV4157-206 double-blind phase results; falls were captured as adverse events if reported as "worsening falls" or if the fall resulted in an injury.

BHV4157-206-RWE: Study Designed In Discussion with FDA

FDA Feedback	BHV4157-206-RWE Protocol
Follow Industry Guidance for RWE*	Regulatory precedent for NDA approval based on RWE
Submit Protocol and Analysis Plan for FDA review prior to database lock	Prespecified endpoints and analysis plan based on FDA input
Use US SCA Natural History cohort as external control for primary analysis	Minimizes potential for bias: Biohaven trial & US SCA Natural History study conducted by same sites/investigators, evaluating similar scales, over similar time period, with same population, on same standard of care treatment
Use Propensity Score Matching (PSM) methodology	Minimizes potential for bias by balancing baseline characteristics between treatment group and external control; Used in other NDAs leveraging RWE**
Match populations based on trinucleotide repeat length	Minimizes potential for bias by matching treatment group and external control based on an additional genetic factor associated with disease burden
Match populations on year 1 progression rates by genotype	Minimizes potential for bias by addressing non-linear patterns of disease progression and inherent heterogeneity of SCA genotypes

*Guidance for Industry Considerations for the Use of Real-World Data and Real-World Evidence to Support Regulatory Decision Making for Drug and Biological Products (https://www.fda.gov/media/171667/download
**Lynch DR, et. al. Propensity matched comparison of omaveloxolone treatment to Friedreich ataxia natural history data. Ann Clin Transl Neurol. 2024 Jan;11(1):4-16. doi: 10.1002/acn3.51897. Epub 2023 Sep 10. PMID: 37691319; PMCID: PMC10791025.

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Expert Perspective on Topline Results

Jeremy Schmahmann, M.D.

Professor of Neurology, Harvard Medical School; Martha and Robert Fogelman Endowed Chair in Ataxia and Cerebellar Ataxia; Director, Mass General Hospital Ataxia Center; and Medical Research Advisory Board, National Ataxia Foundation

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Large safety database demonstrates troriluzole is well tolerated in SCA

"I Cannot Underscore Enough the Impact of a Potential Treatment That Can Slow SCA Disease Progression"

SCA Commercial Planning

John Tilton
Chief Commercial Officer, Rare Disease, Biohaven

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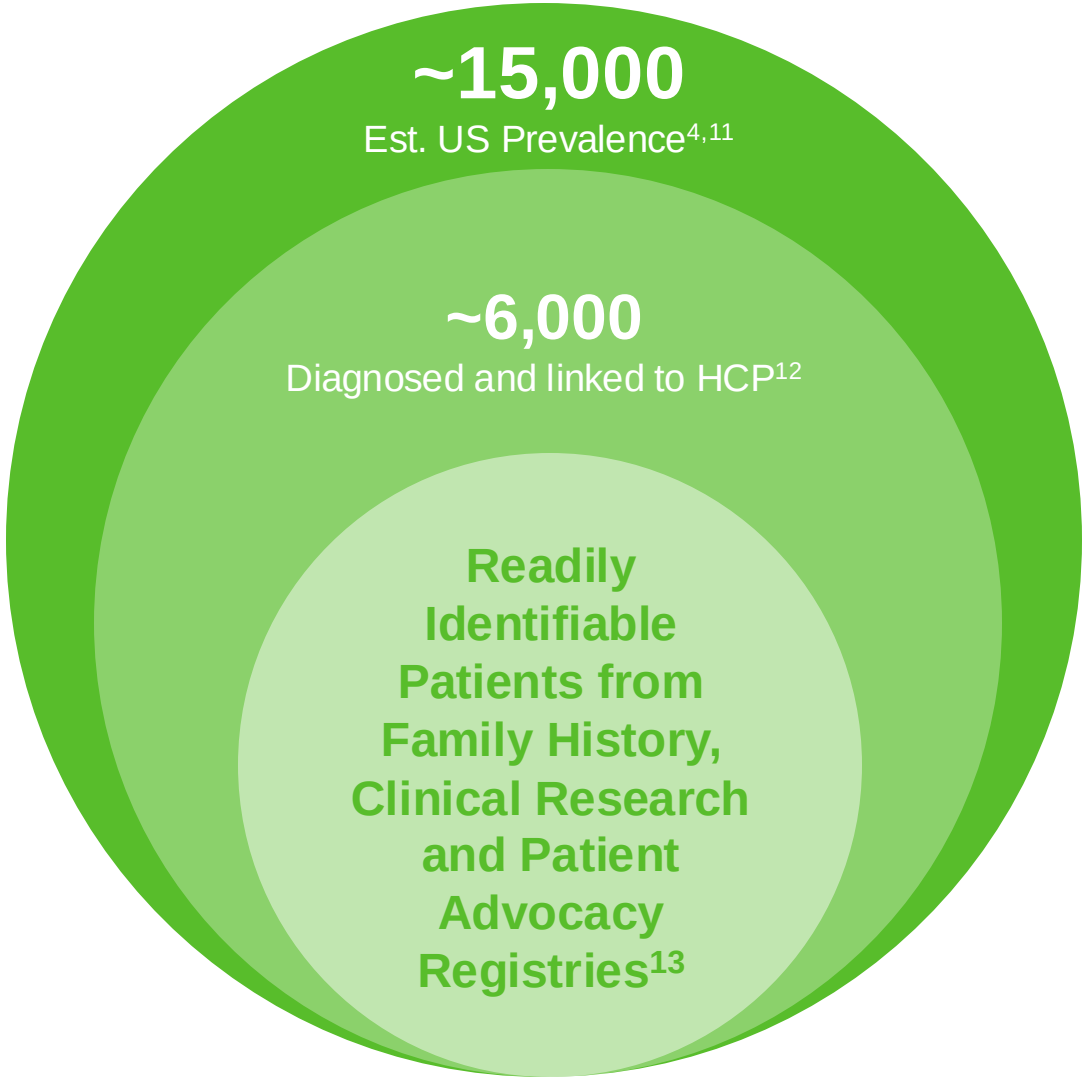
SCA Represents a Significant Commercial Opportunity



- ~6,000 diagnosed US patients
- No currently approved SCA treatments
- Availability of genetic testing and advent of approved treatment will facilitate diagnosis
- Engaged, connected SCA patient community
- Strong patient advocacy support
- KOLs, HCPs and key centers treating SCA have been identified

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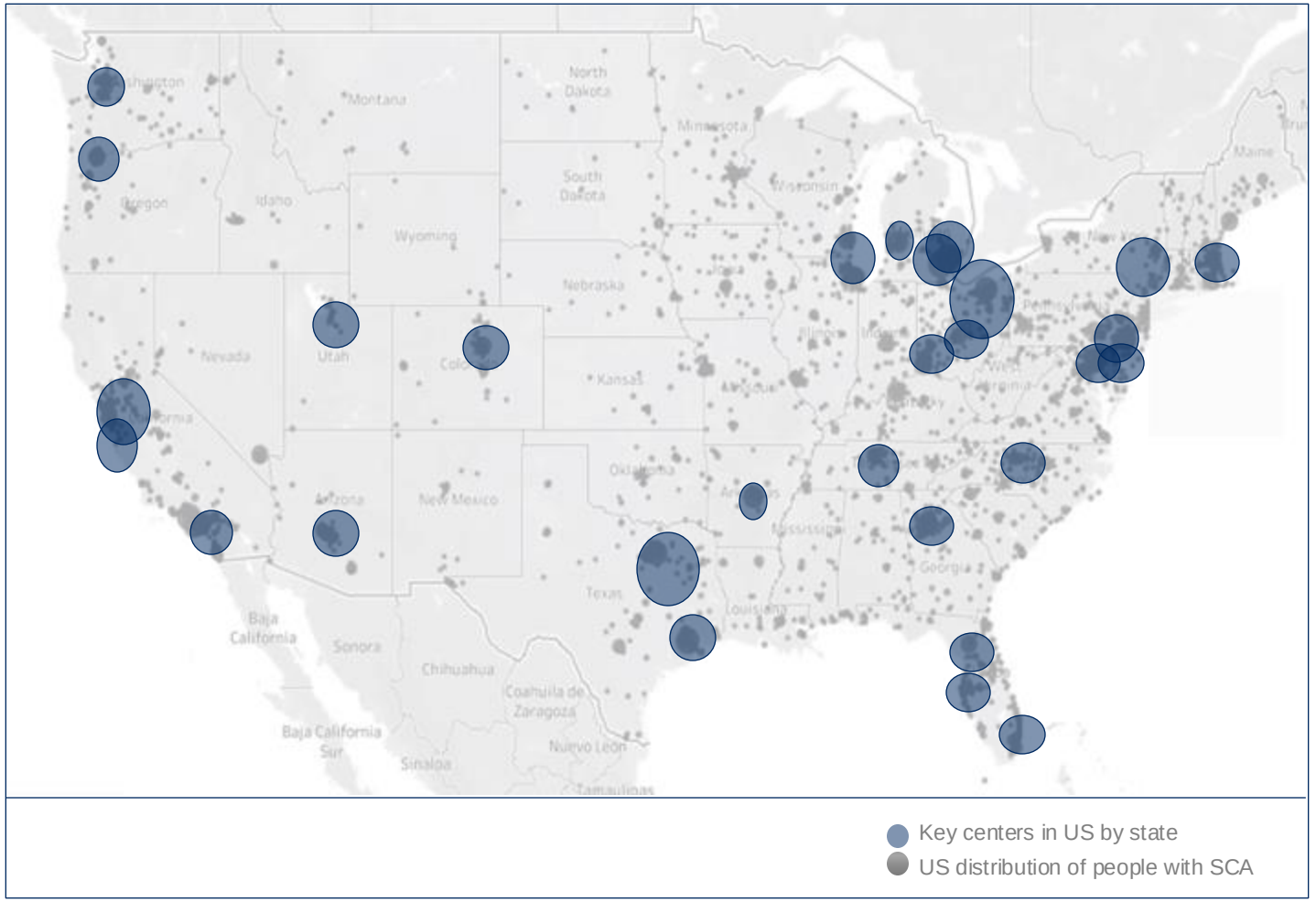
Significant Commercial Opportunity: SCA Centralized Treatment Allows for Targeted and Efficient Commercialization Plan

SCA treatment at key centers

121 KOLs, 22 NAF Ataxia Centers of Excellence and 73 additional Movement Disorder and Ataxia Centers have been identified and manage many patients with SCA[11,13]

Experienced, efficient commercial team

Commercial team of ~50 staff will drive a focused and rapid troriluzole launch



Experienced Commercial Team Actively Preparing for Anticipated Launch

SCA Launch Priorities

PRIORITY 1
Identify patients, drive early diagnosis

PRIORITY 2
Establish troriluzole as SoC in SCA

PRIORITY 3
Create access and reimbursement

PRIORITY 4
Ensure ongoing treatment

Launch preparation has accelerated with planned 4Q 2024 NDA submission

Our commercial team has a proven track record in successful rare disease launches

Commercial strategies and tactical plans have been developed

The Biohaven team will be prepared to serve patients living with SCA if approved

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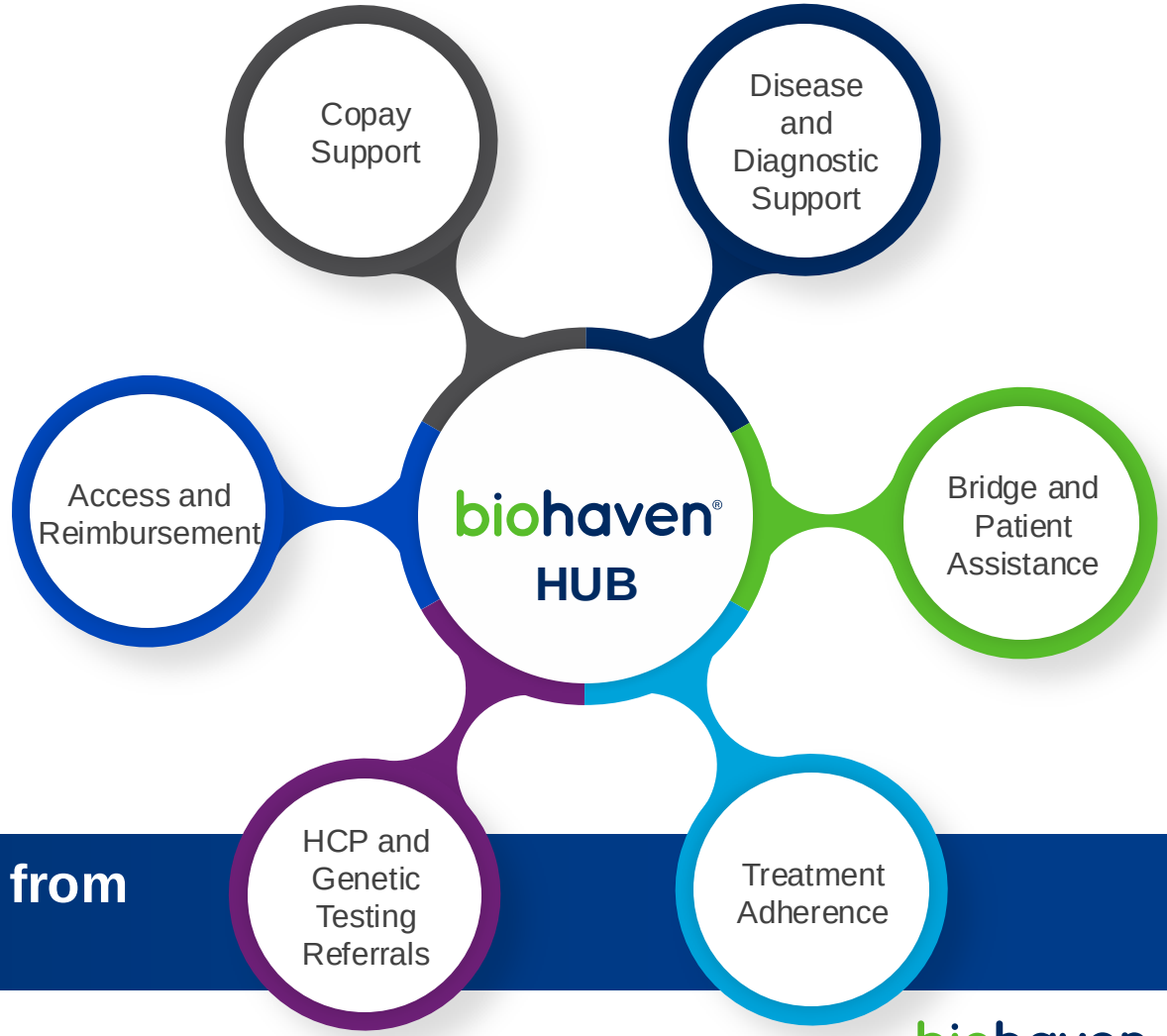
Comprehensive Patient Access and HCP Support

Biohaven HUB will be a central point of contact for patients, caregivers and HCPs

- Process new patient starts and facilitate bridge program
- Insurance and reimbursement support
- Copay, early access and patient assistance program
- Integrated specialty pharmacy
- Highly coordinated field reimbursement and case management teams
- Disease education and diagnostic assistance
- Treatment adherence program



KEY POINT

All SCA patients who can benefit from troriluzole should have access

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Biohaven Collaborates with Ataxia Organizations to Serve Patients

- **COLLABORATIONS** with leading SCA researchers
- **PARTNERSHIPS** with advocacy organizations
- **SCIENTIFIC MEMBERSHIP** in research groups
- **KEY CONTRIBUTION** to the ongoing development of a quality natural history study in SCA

National Ataxia Foundation

Biohaven Pharmaceuticals Inaugural Partner of National Ataxia Foundation's Drug Development Collaborative

ARCA GLOBAL

MGH 1811

EBERHARD KARLS UNIVERSITÄT TÜBINGEN

CPTA CRITICAL PATH TO THERAPEUTICS FOR THE ATAXIAS

CRC-SCA Clinical Research Consortium for the Study of Cerebellar Ataxia

ATAXIA Ataxia UK

HOUSTON Methodist CONTINUING CARE HOSPITAL

ATAXIA GLOBAL INITIATIVE

SCA Global

MDA Muscular Dystrophy Association

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Q&A Session

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Thank You!

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