

# Real-World Strategies to Enhance the Palatability of Coformulated Sodium Phenylbutyrate and Taurursodiol for the Treatment of Amyotrophic Lateral Sclerosis

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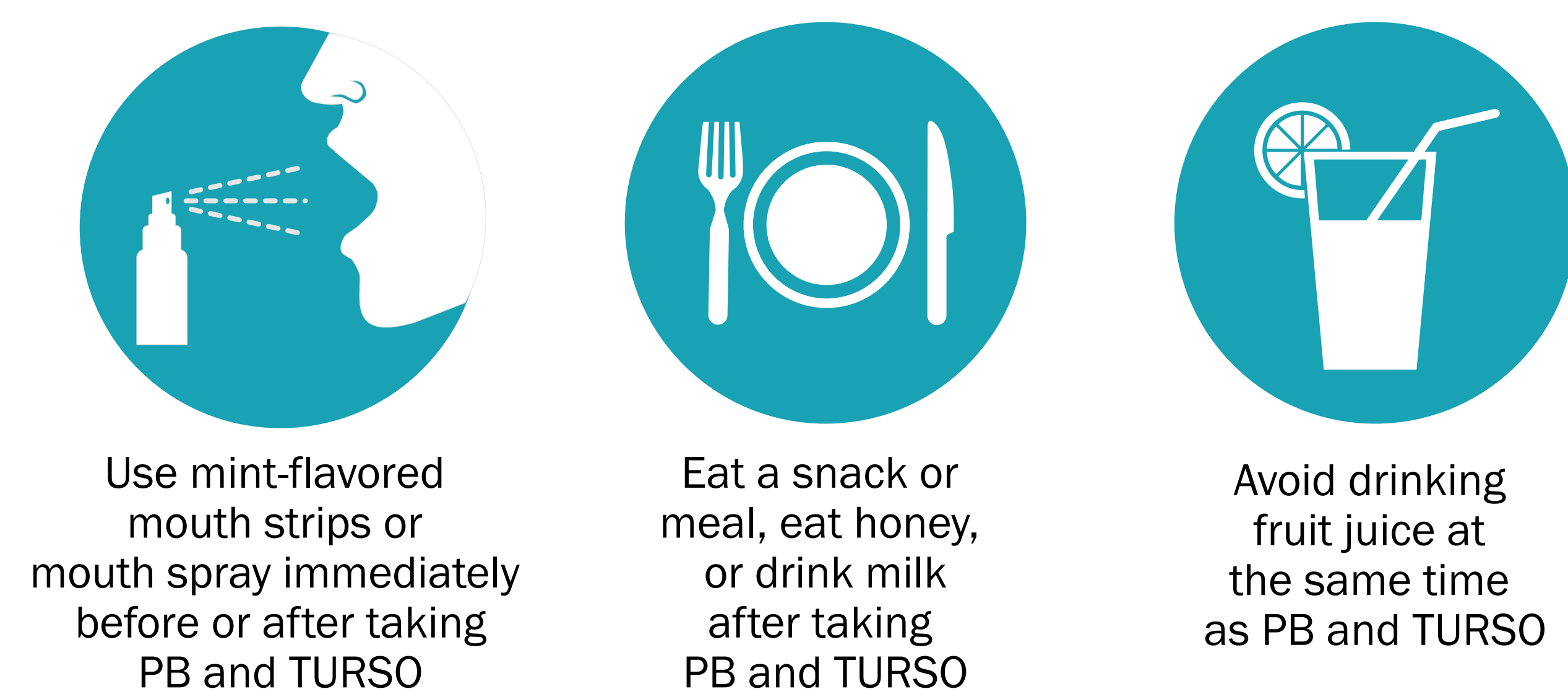
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## BACKGROUND

- An oral, fixed-dose combination of sodium phenylbutyrate and taurursodiol/ursodoxicoltaurine (PB and TURSO) is approved to treat amyotrophic lateral sclerosis (ALS) in adults in the United States<sup>1</sup> and approved with conditions to treat ALS in Canada<sup>2</sup>
- PB and TURSO is a powder administered orally or via feeding tube after suspension in room temperature water.<sup>1,2</sup> PB and TURSO can have a bitter taste to some people living with ALS (PLWALS) who take it orally, which may compromise adherence and, consequently, treatment effectiveness
- The Canadian product monograph provides specific details around how PLWALS can reduce the bitter aftertaste (Figure 1)<sup>2</sup>

Figure 1. Methods for Reducing Bitter Aftertaste of PB and TURSO<sup>2</sup>



Per Canadian product monograph.<sup>2</sup> PB and TURSO, sodium phenylbutyrate and taurursodiol.

## TASTE IN PLWALS

- PLWALS may have altered taste perception for a multitude of reasons, including degeneration of the solitary nuclei, impairment of the cranial nerves innervating the taste buds, medications, and genetics<sup>3,4</sup>
- In particular, differential disease effects on the cranial nerves may preserve or potentiate output from gustatory cells located in the circumvallate papillae that perceive bitter taste<sup>4,5</sup>

- Prior to marketing approval in Canada, PB and TURSO was available through the Health Canada Special Access Program (SAP). As of September 13, 2022, 272 PLWALS with a mean age of 63 years received PB and TURSO through the SAP

**PB and TURSO is an investigational drug not approved for use pending regulatory review in the European Medicines Agency for the treatment of ALS.**



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## OBJECTIVE

- To explore real-world coping strategies of PLWALS and health care practitioners for potentially enhancing PB and TURSO palatability based on their experiences as part of the PB and TURSO Canadian SAP

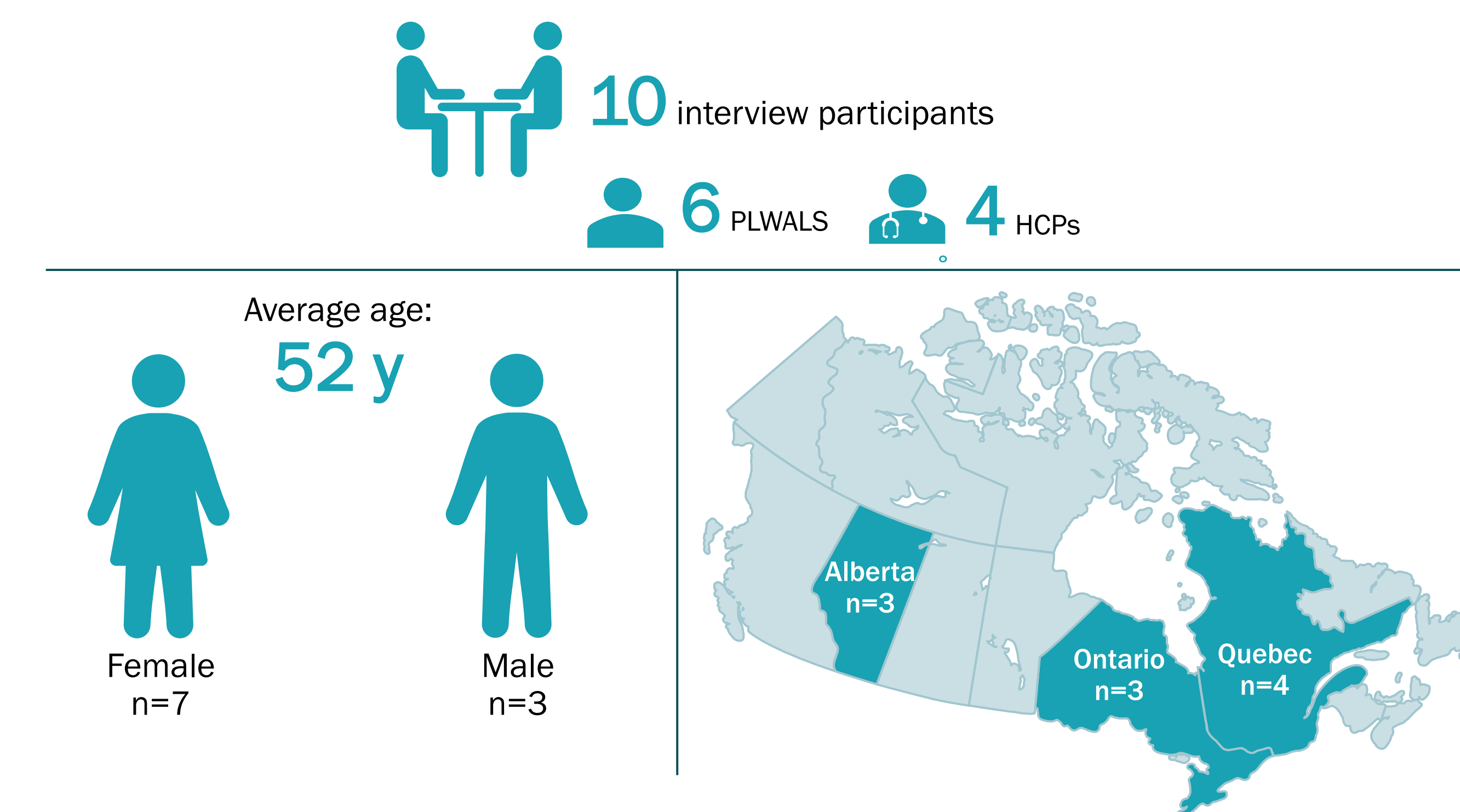
## METHODS

- One registered dietitian conducted individual, semistructured qualitative interviews with PLWALS receiving PB and TURSO via oral administration (n=6), ALS clinic nurses/coordinators (n=3), and a neurologist specializing in ALS care (n=1)
- Participant responses were transcribed, and data were analyzed to generate themes

## RESULTS

Participant characteristics are summarized in (Figure 2)

Figure 2. Participant Characteristics



HCP, health care practitioner; PLWALS, people living with amyotrophic lateral sclerosis.

- All PLWALS who responded (100%) reported using  $\geq 1$  of the coping strategies identified
- None of the PLWALS (0%) reported discontinuing treatment owing to taste
  - For context, of the 217 Canadian PLWALS exposed to PB and TURSO treatment through the SAP, 8 (3.7%) cited bitter taste among their reasons for discontinuation

Multiple real-world coping strategies to mask the bitter taste of PB and TURSO were identified in addition to those recommended in the Canadian product monograph for PB and TURSO. Altogether, these strategies encompassed 3 themes (Table 1):

- Optimism and hope in chronic disease
- Medication administration techniques
- Taste-enhancing techniques

Table 1. Real-World Coping Strategies for Enhancing PB and TURSO Palatability

Optimism and Hope in Chronic Disease	Medication Administration Techniques	Taste-Enhancing Techniques
<ul style="list-style-type: none"> <li>Feelings of hope help outweigh bitter taste of the therapy</li> <li>Ability to accept the taste and the potential positive impact of treatment on disease symptoms</li> <li>Taste buds have changed with the disease, so bitter taste is less of an issue</li> <li>Conversations (verbal/email) with HCPs about how to optimize medication palatability are very helpful with medication adherence</li> </ul>	<ul style="list-style-type: none"> <li>Mix medication with a milk frother<sup>a</sup></li> <li>Shake suspension in a mason jar<sup>a</sup></li> <li>Stir suspension with a spoon</li> <li>Drink suspension with a straw</li> <li>Pinch nose and drink suspension<sup>a</sup></li> <li>Drink suspension in one gulp<sup>a</sup></li> </ul>	<ul style="list-style-type: none"> <li>After taking the suspension, try Nature's Wild Berry<sup>a</sup></li> <li>After taking the suspension, try chocolate pudding, chocolate, a banana, a boiled egg, salt-and-vinegar potato chips, crackers, cookies, applesauce, black coffee, yogurt, Listerine strips, or mouthwash</li> <li>After taking the suspension, brush teeth<sup>a</sup></li> <li>Avoid eating or drinking anything citrusy after taking the suspension, as that makes the taste more bitter</li> </ul>

<sup>a</sup>Denotes strategies that are not included in the Canadian product monograph. HCP, health care practitioner; PB and TURSO, sodium phenylbutyrate and taurursodiol.

## CONCLUSION

Although further studies are needed to substantiate and enrich these preliminary results, these data suggest that a variety of strategies may be successfully applied in the real-world setting to manage taste expectations and potentially enhance the palatability of PB and TURSO

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### Disclosures

NS was consulted by Amylyx to conduct the interviews. SW, TJ, and JT are full-time employees of Amylyx.

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