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## Abstract

**Background** Hereditary Angioedema (HAE) is a complex debilitating disease that is often misdiagnosed and under treated. Our study objective was to gain insight into which treatments Canadians are using to treat their acute attacks, prophylactic treatments, and their frequency of administration.

**Methods** In 2017-2018, data was collected through voluntary online surveys of children, youth, and adults who live with HAE and their caregivers in Canada. The following data was based solely on adult participants.

**Results** Our Canadian participants are using the following to treat attacks: C1 esterase Inhibitor (Berinert®)-61%, C1 esterase Inhibitor (Cinryze®)-3%, Icatibant (Firazyr®)-10%, Androgen (Danazol®)-5%, Tranexamic acid-3%, No pharmaceutical treatment-3%, and Other-15%. Berinert is being used by patients: on demand (36%), chronic prophylaxis only (9%), chronic prophylaxis and on demand (55%). The majority of respondents use Berinert weekly or twice weekly. Cinryze is being administered as: chronic prophylaxis only (33%), and for both chronic prophylaxis and on demand (67%). All respondents equally treat at different intervals. Firazyr is being administered as: on demand (89%), and for both chronic prophylaxis and on demand (11%). Half of the participants require only one treatment for an acute attack and rarely use it for prophylaxis. Danazol is used for: on demand (25%), chronic prophylaxis only (25%), chronic prophylaxis and on demand (50%). All respondents equally treat at different intervals. Tranexamic acid is being taken for chronic prophylaxis and on demand (100%) by all respondents.

**Conclusion** The data collected demonstrates that treatments are being used interchangeably for acute and prophylaxis treatment despite the indications listed on the product monographs. These results validate that patients in consultation with their HAE specialists have determined an individualized treatment schedule that works best to control the symptoms of their disease. All results are limited to the respondents and may not represent the broader Canadian HAE population

## Introduction

- HAE is a debilitating and unpredictable disease that can be controlled using various treatment options.
- The available medications are used for treatment of on-demand acute attacks, chronic prophylaxis to prevent attacks or both.
- The available medications vary in treatment frequency, dose and route of administration
- We sought to better understand the choice of treatment and the ways in which these are being used in Canadian HAE patients.

## Methods

Starting in January 2017, all members of HAE Canada were emailed an extensive survey to gather data on patients living with HAE and their caretakers. This was the first HAE national survey done in Canada, being referred to as the "Report Card on HAE in Canada". Data was gathered and analyzed in October 2017 by using the percentage of total number of surveys answered for each question.

## Results

### Primary Choice Of Medication Used By Canadian HAE Patients

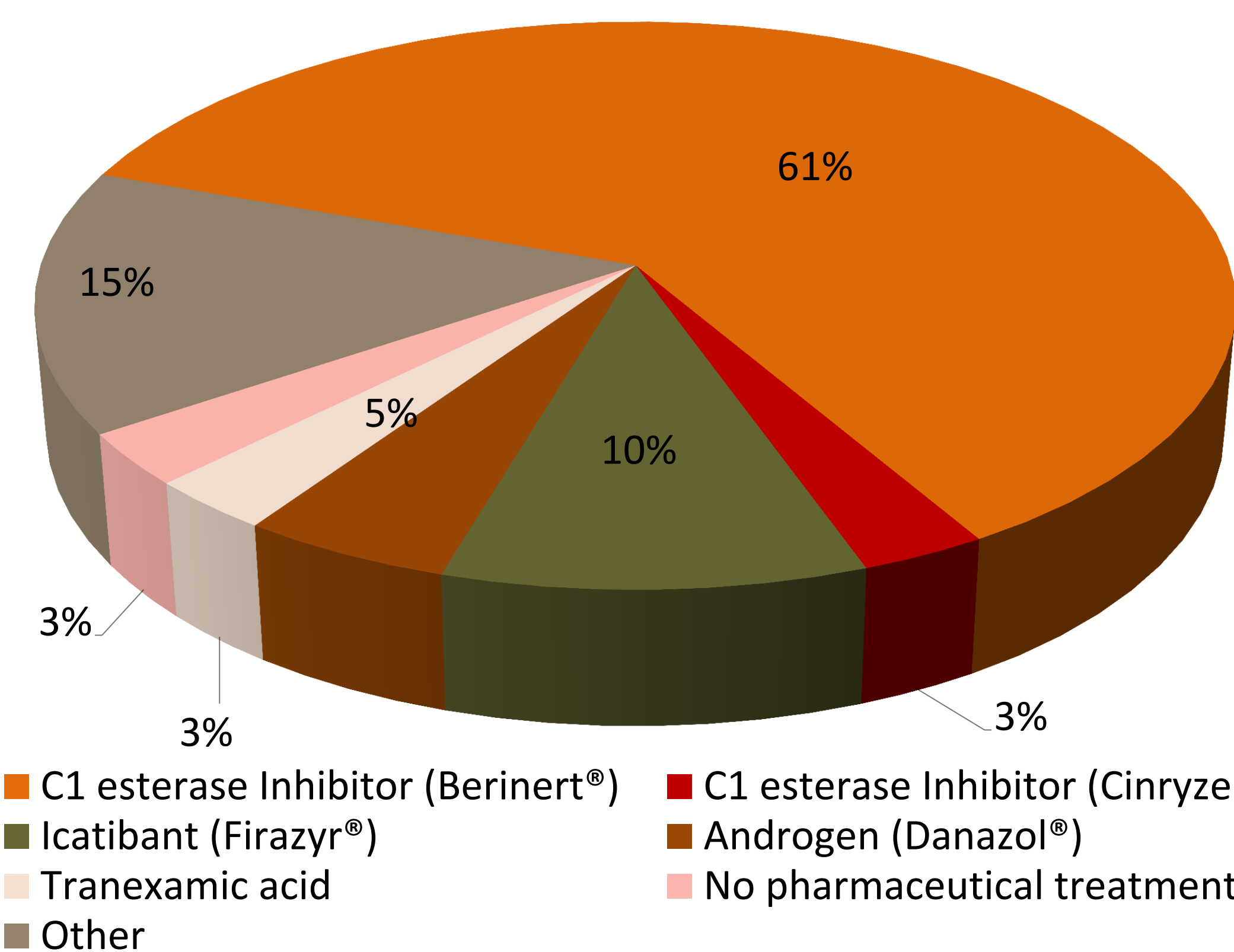


Figure 1: N= 87

### Regular Treatments Of Berinert® By Canadian HAE Patients

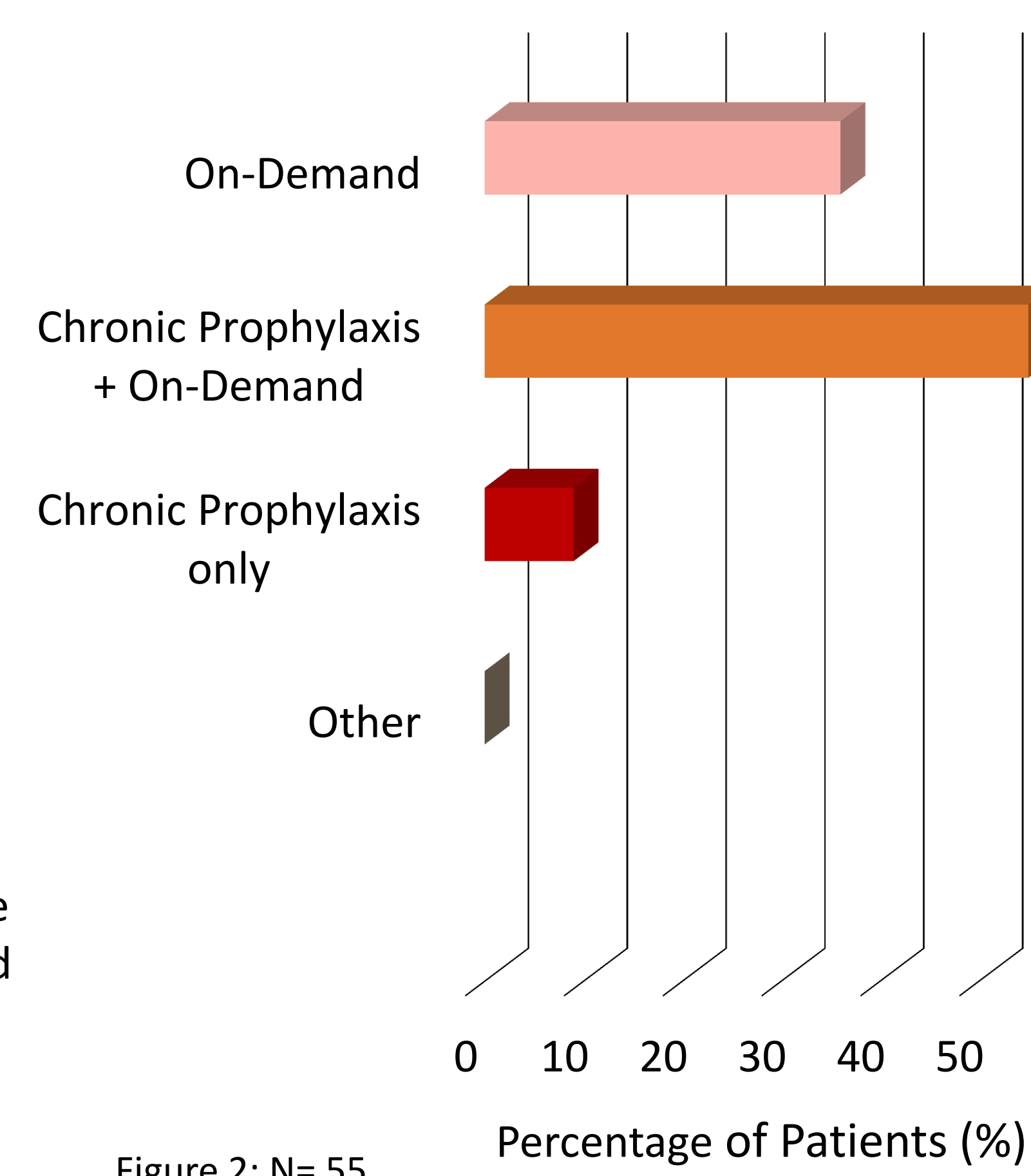


Figure 2: N= 55

### Usage Patterns Of Berinert® By Canadian HAE Patients

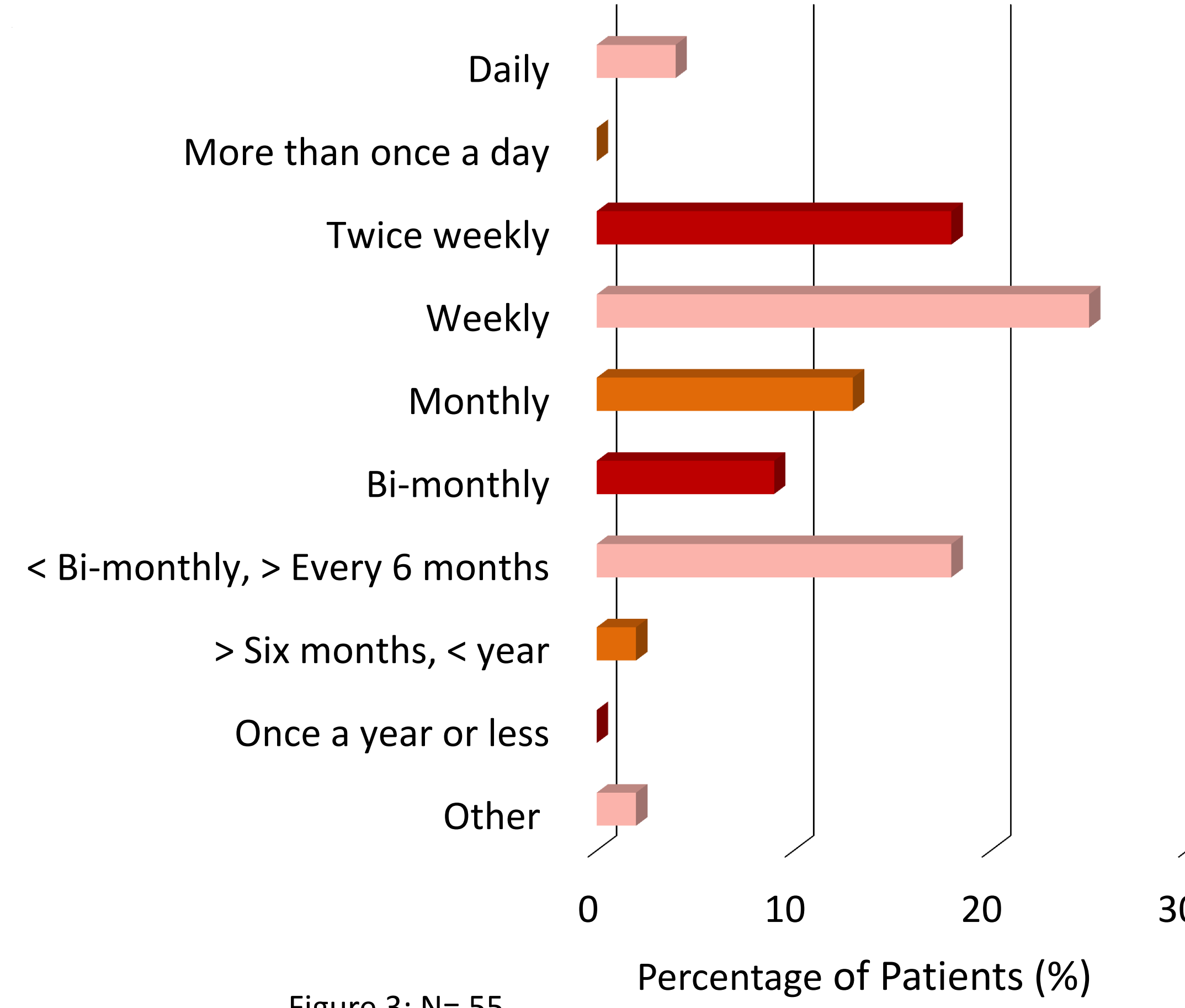


Figure 3: N= 55

### Regular Treatment Of Danazol® By Canadian HAE Patients

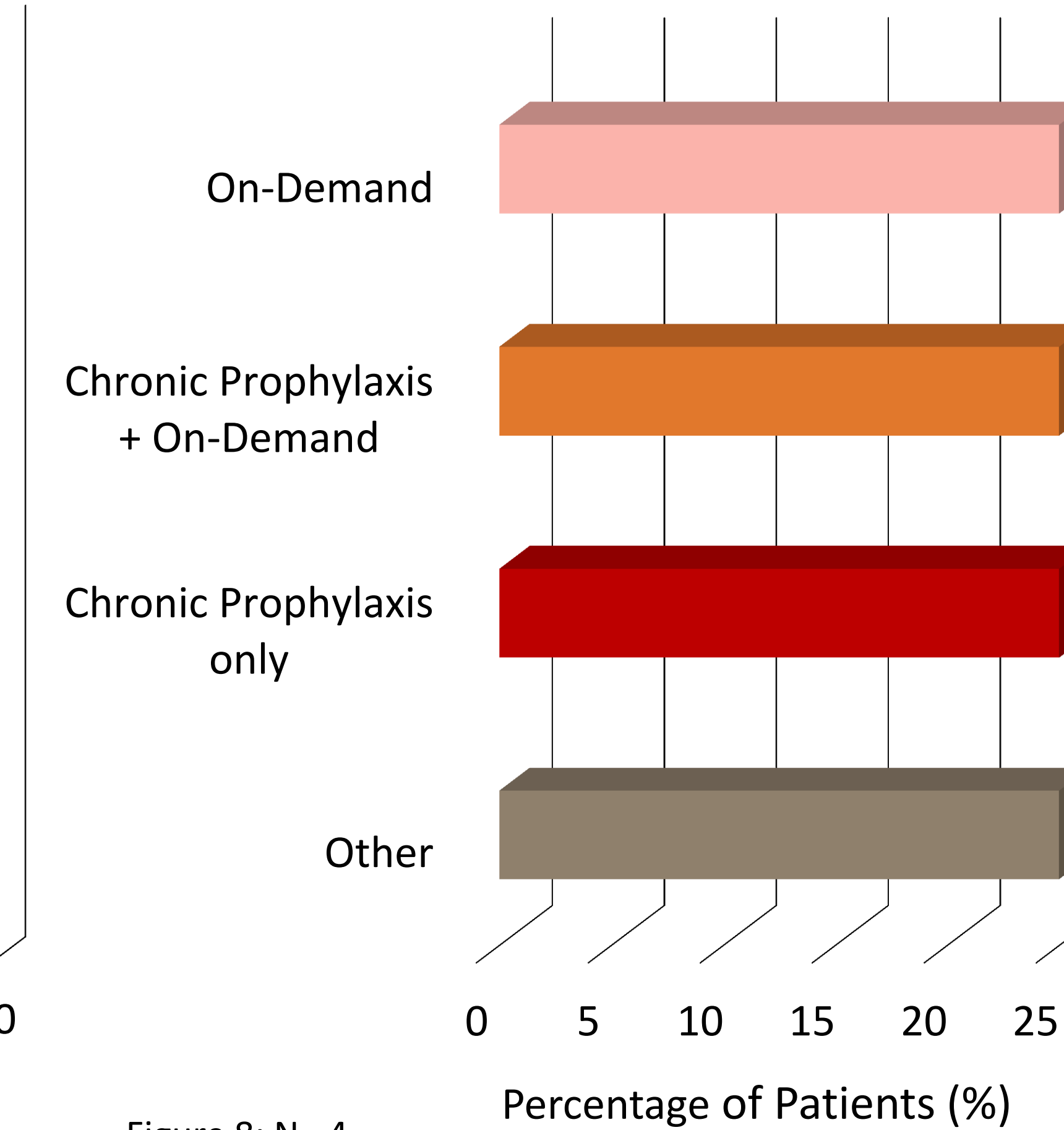


Figure 8: N= 4

### Usage Patterns Of Danazol® By Canadian HAE Patients

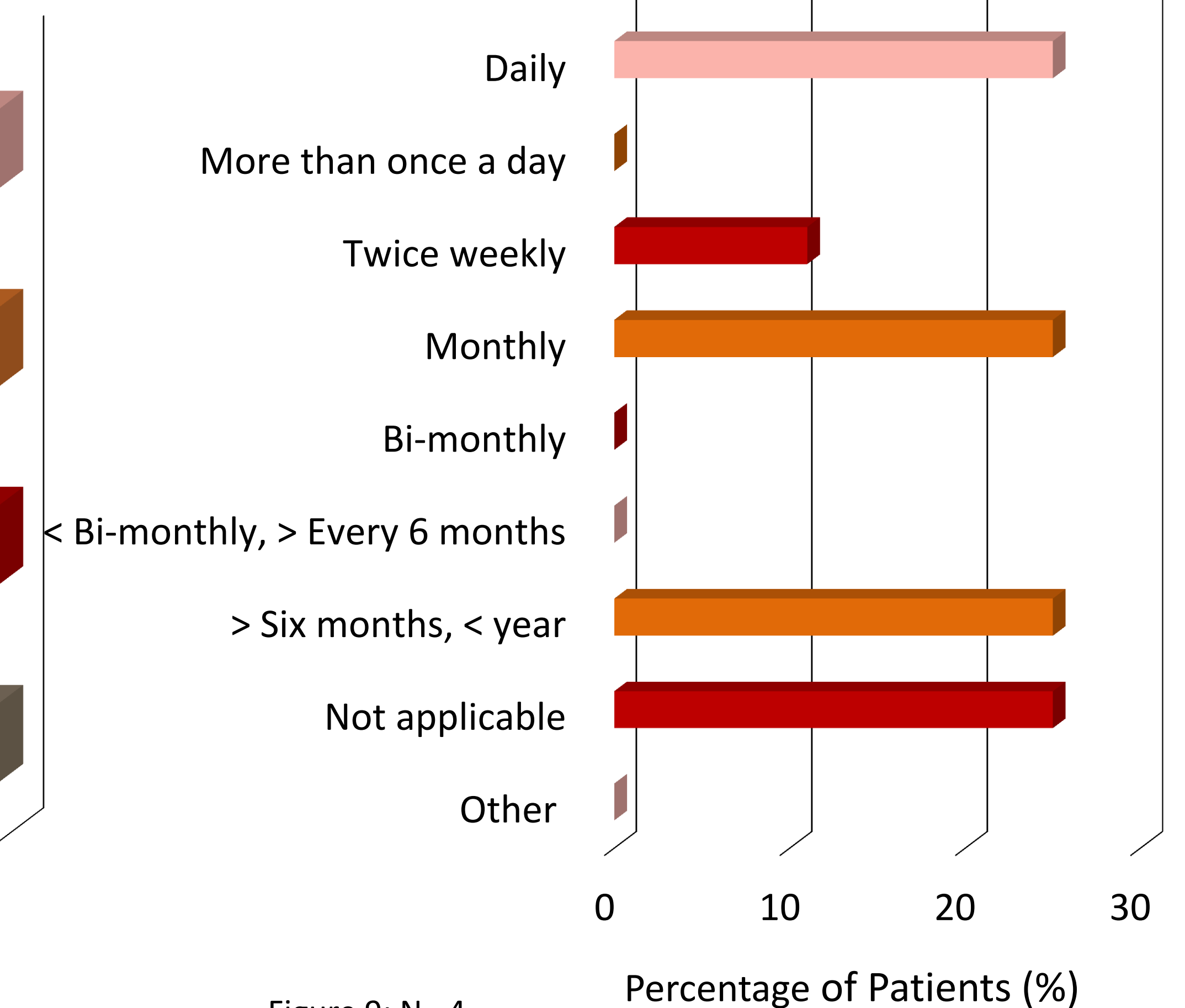


Figure 9: N= 4

### Regular Treatments Of Cinryze® By Canadian HAE Patients

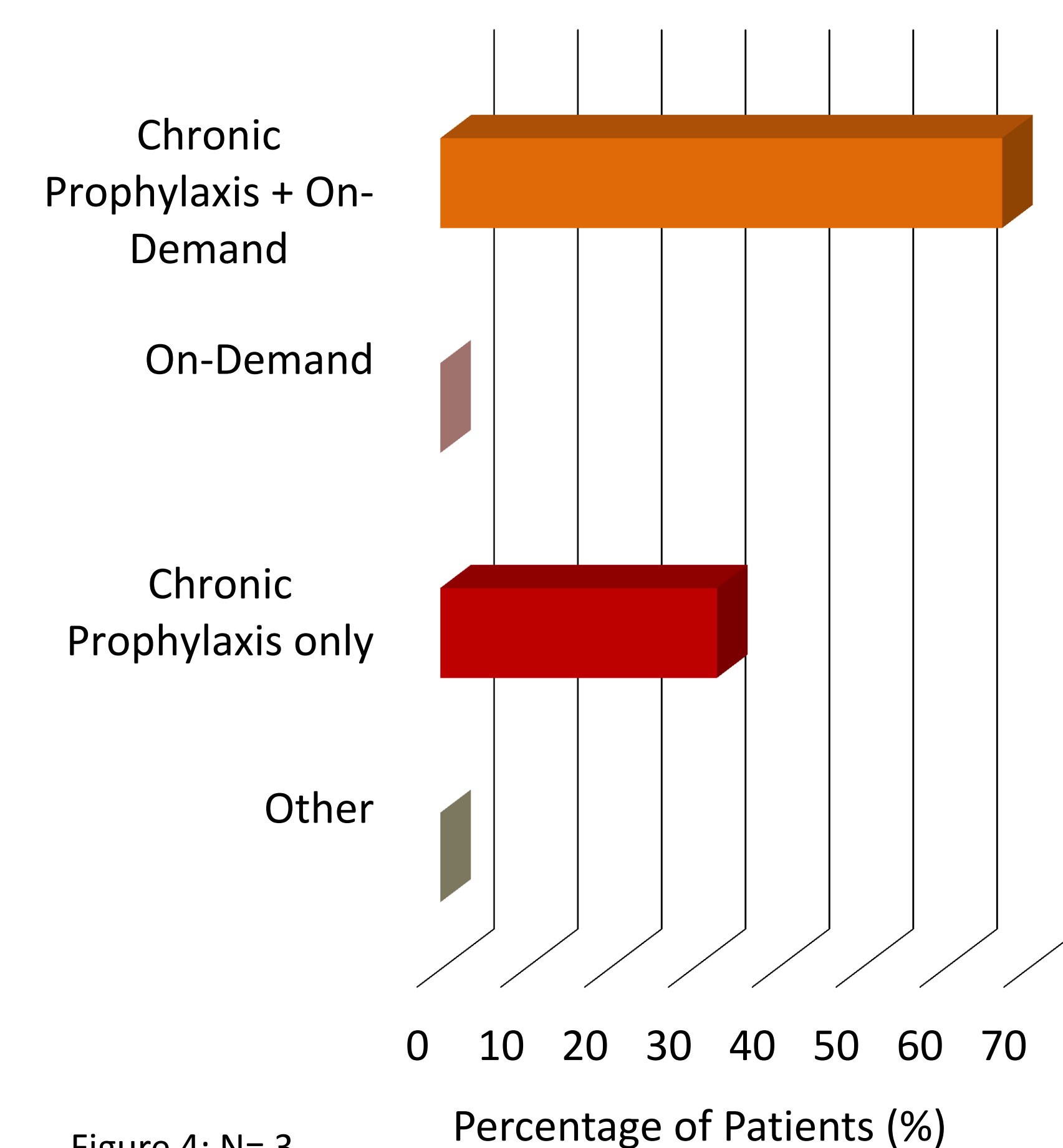


Figure 4: N= 3

### Usage Patterns Of Cinryze® By Canadian HAE Patients

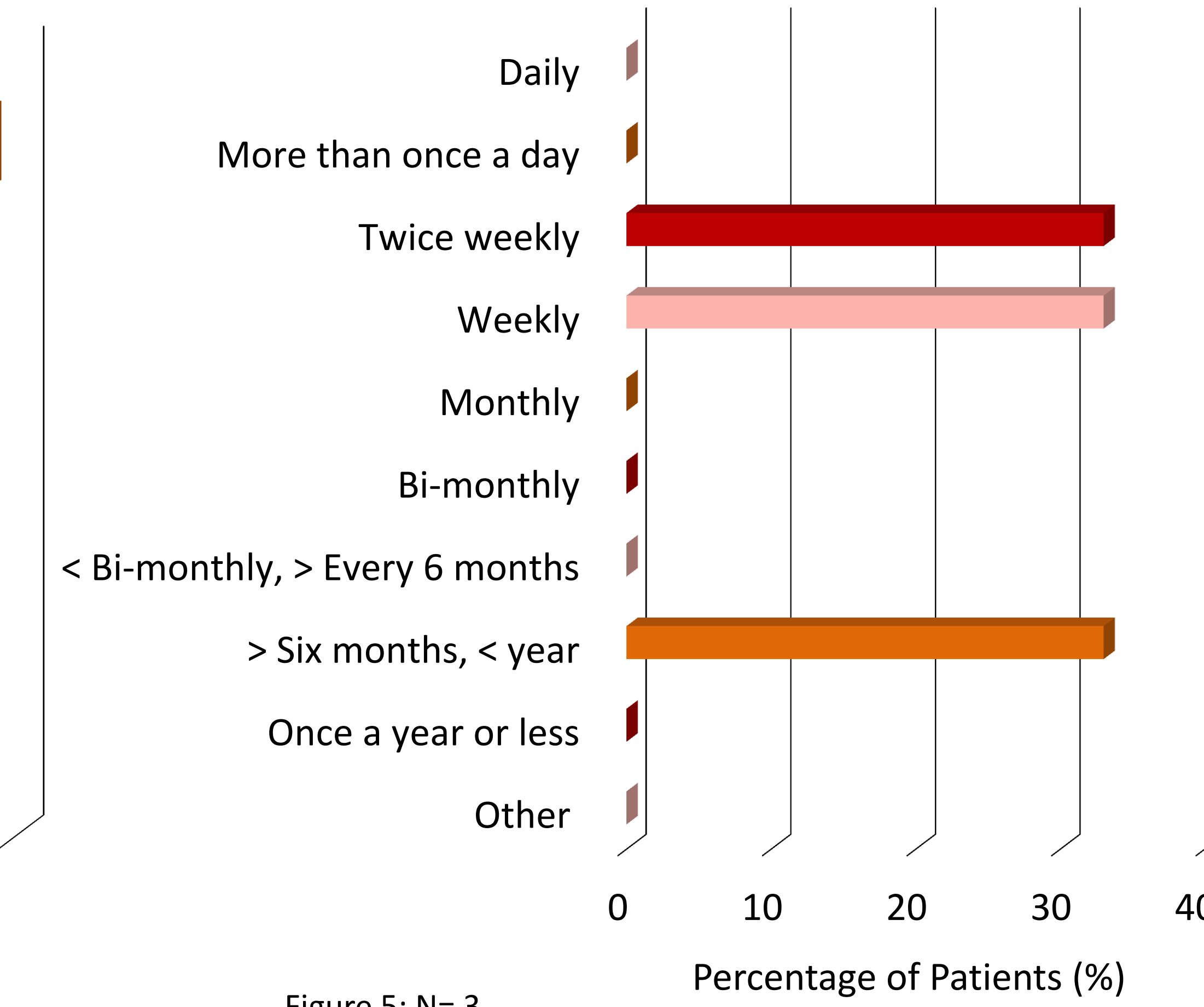


Figure 5: N= 3

### Regular Treatment Of Tranexamic Acid By Canadian HAE Patients

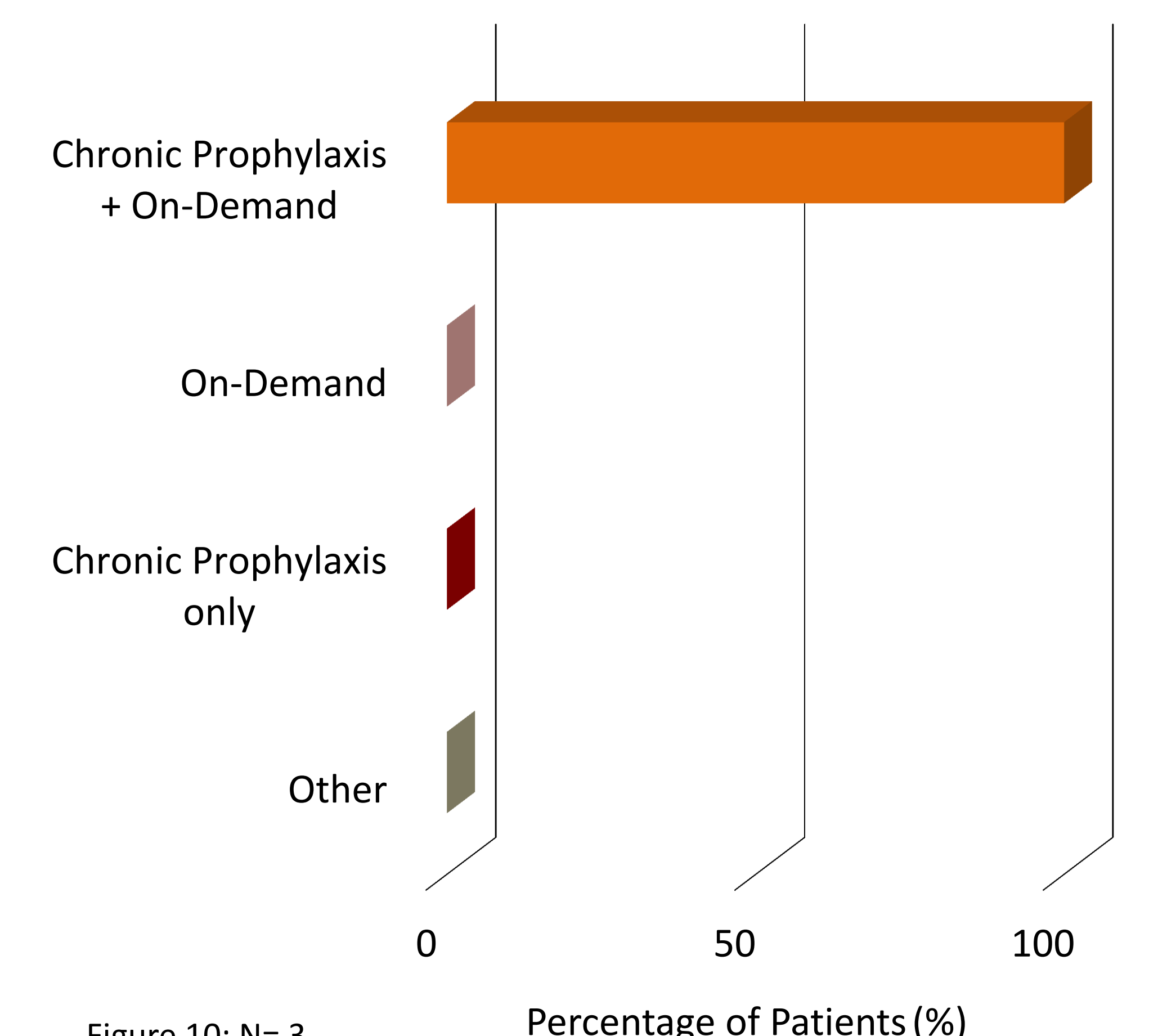


Figure 10: N= 3

### Regular Treatment Of Firazyr® By Canadian HAE Patients

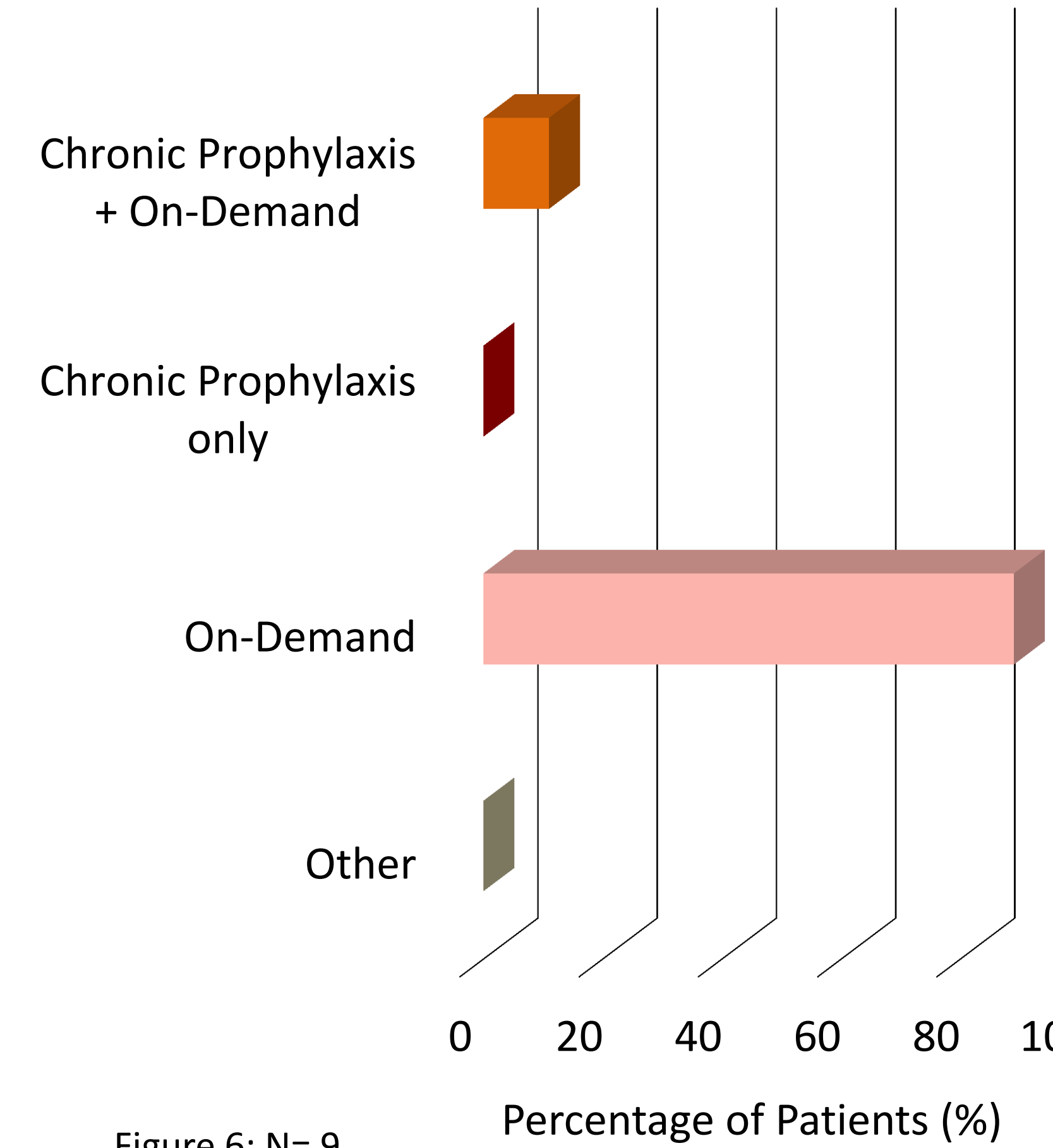


Figure 6: N= 9

### Usage Patterns Of Firazyr® By Canadian HAE Patients

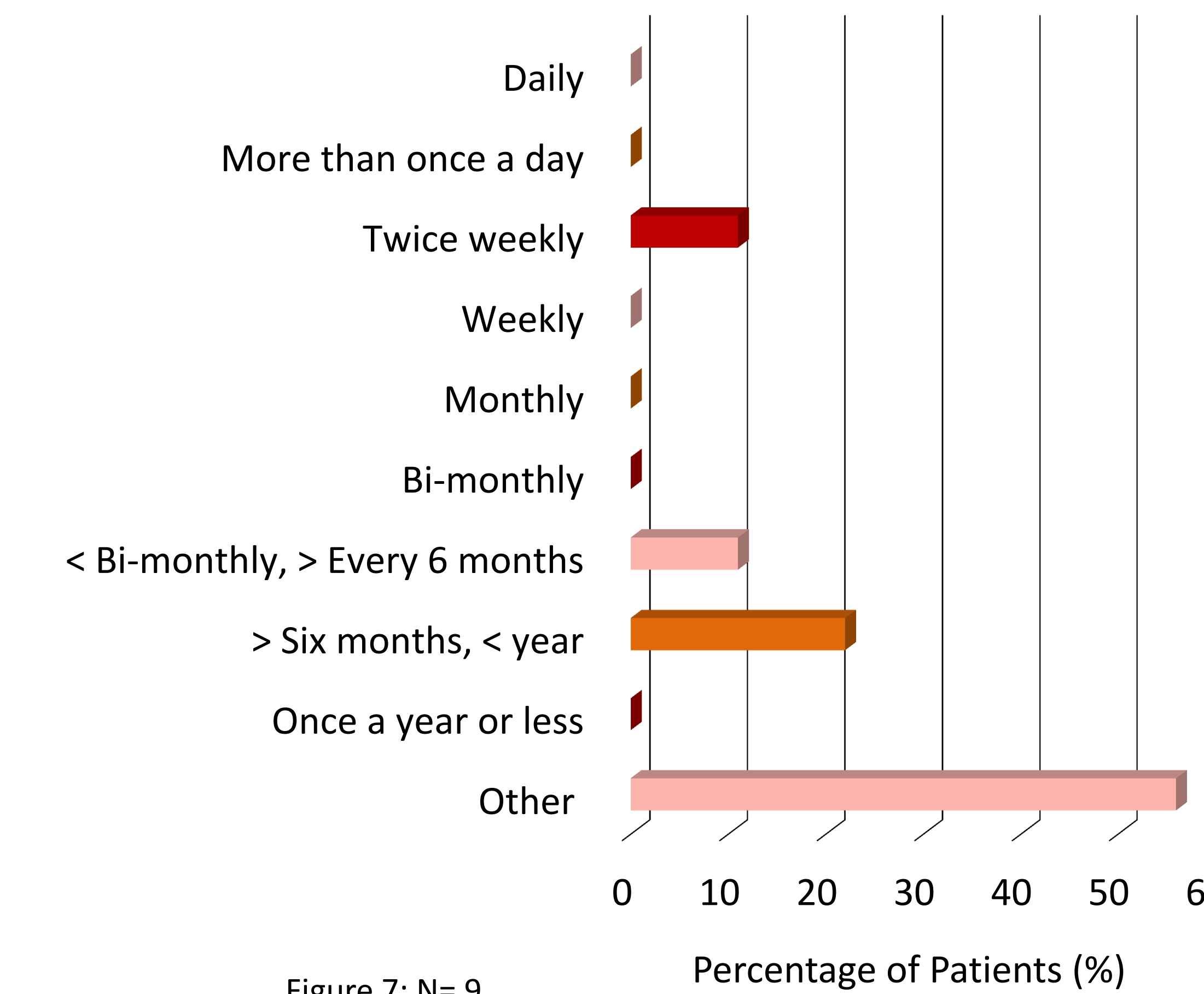


Figure 7: N= 9

## Conclusion

HAE patients in Canada are using treatments to treat acute attacks on-demand and to prevent attacks by chronic prophylaxis. The frequency of their administration differs greatly between each medication. These results may be used to compare treatment usage patterns in other countries.

## References

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2. Banerji, A., Busse, P., Shennak, M., Lumry, W., Davis-Lorton, M., et al (2017). Inhibiting Plasma Kallikrein for Hereditary Angioedema Prophylaxis. *New England Journal of Medicine*, 376(8), 717-728. doi: 10.1056/NEJMoa1605767

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