

The Humanistic and Economic Impact of Fibrodysplasia Ossificans Progressiva on Primary Caregivers: Results from an International Burden of Illness Survey

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Background

- Fibrodysplasia ossificans progressiva (FOP; OMIM #135100) is an ultra-rare, genetic disorder characterised by heterotopic ossification in soft and connective tissues causing severe, irreversible disability.^{1,2}
- Disability in FOP is cumulative; most people with the disorder use a wheelchair by their third decade of life and require lifelong assistance in performing activities of daily living.^{3,4}
- While recent research has enhanced understanding of the clinical aspects of FOP, less is known about the humanistic and economic impact of FOP on primary caregivers.

Objective

To determine the social, emotional, health/psychological, and employment impact of supporting a family member with FOP on primary caregivers

Methods

- An international, cross-sectional burden of illness survey (NCT04665323) was co-created with advisors from the FOP community.
- The survey was available online between 18th January and 30th April 2021 across 15 countries and in 11 languages.
- Participants were recruited through the International FOP Association (IFOPA) and national/regional FOP organisations.
- People with FOP of any age, as well as their immediate family members (parents/legal guardians and siblings aged ≥18 years), were eligible to participate;
 - Family members who identified as the primary caregiver for the person with FOP answered additional survey questions.
- All family members completed a bespoke, FOP-specific questionnaire that assessed the social and emotional impact of caring for a family member with FOP.
- Primary caregivers completed the Zarit Burden Interview (ZBI) that assessed the impact of caring for a family member with FOP on health and/or psychological wellbeing:
 - ZBI total scores range from 0–88 and impact was assessed as the following: <21, little/absent; 21–40, mild to moderate; 41–60, moderate to severe; 61–88, severe.
- All family members answered questions related to the impact of FOP on their careers.

Results

- 163 primary caregivers responded to the survey:
 - Mean (standard deviation [SD]) age of primary caregivers was 49.2 (12.0) years and 85.4% identified as female;
 - The number of primary caregiver responses by country is presented in **Figure 1**.
- Primary caregivers reported a large emotional impact of supporting their family member with FOP (**Figures 2A, 2B, 2C**):
 - Additionally, most primary caregivers often or always found it emotionally difficult to have a family member with FOP and worried about their family member with FOP hurting themselves (**Figures 2D, 2E**);
 - Over two-thirds of primary caregivers reported that caring for their family member with FOP had little or no impact on their social interactions (**Figure 2F**).
- The activities for which primary caregivers most often assisted their family member with FOP are presented in **Figure 3**:
 - Primary caregivers spent a mean (SD) of 10.6 (8.6) hours per day looking after their family member with FOP.
- Close to half (45.1%) of primary caregivers reported a ZBI total score indicating that supporting their family member with FOP had a mild to moderate impact on their health and/or psychological wellbeing:
 - Fewer primary caregivers reported a ZBI total score indicating little/absent (39.9%), moderate to severe (13.7%) or severe (1.3%) impact.
- Primary caregivers supporting younger people with FOP (<8 years) reported a greater impact of caregiving on their health and/or psychological wellbeing compared with those supporting older people with FOP (≥25 years; **Figure 4**).
- Over half of primary caregivers (51.3%) felt they needed to adapt their career to support their family member with FOP.

CONCLUSIONS

- Primary caregivers of people with FOP reported experiencing a wide range of humanistic impacts.
- Experiences such as learning of their family member's FOP diagnosis and supporting their family member through flare-up symptoms were reported to be emotionally challenging by the majority of primary caregivers.
- Most primary caregivers reported needing to adapt their careers to support their family member with FOP.
- These data highlight the personal and societal impacts of caring for an individual with FOP and may help to identify areas of unmet need and inform additional support strategies for the FOP community.

Abbreviations FOP: fibrodysplasia ossificans progressiva; IFOPA: International FOP Association; SD: standard deviation; U.K.: United Kingdom; U.S.: United States; ZBI: Zarit Burden Interview.
References 1. OMIM #135100 FOP. Volume 2020; 2. Kaplan FS et al. J Bone Joint Surg Am 1993;75:220–30; 3. Connor JM & Evans DA. J Bone Joint Surg Br 1982;64:76–83; 4. Pignolo RJ et al. Bone 2020;134:115274.

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Figure 1. Number of primary caregiver responses by country

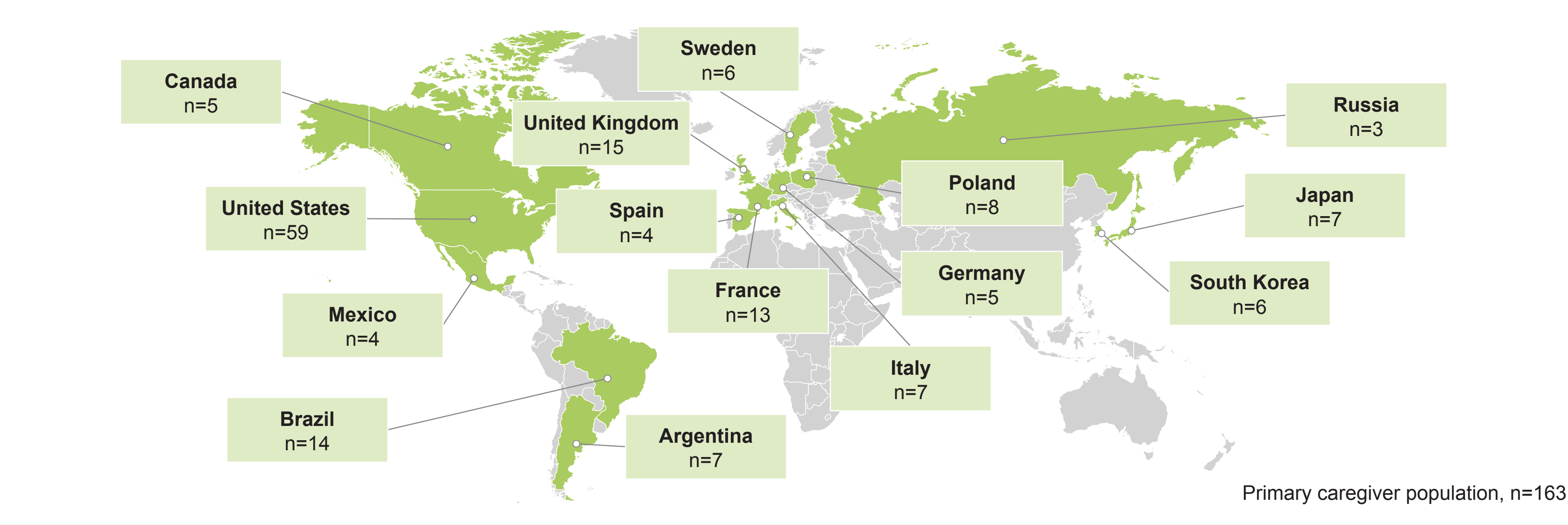


Figure 2. Emotional impact of caring for a family member with FOP on primary caregivers

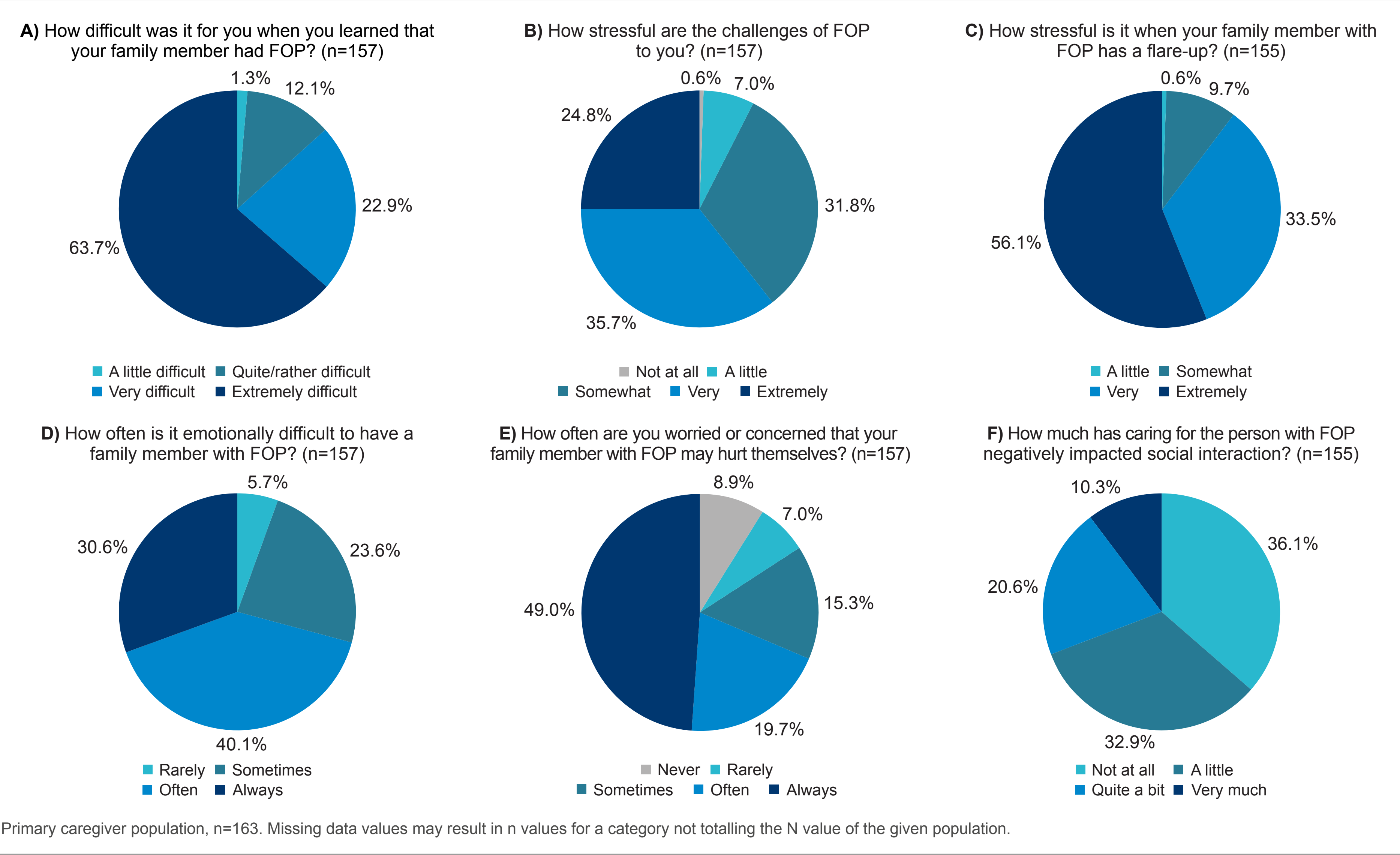


Figure 3. Daily activities for which primary caregivers most often assisted their family member with FOP

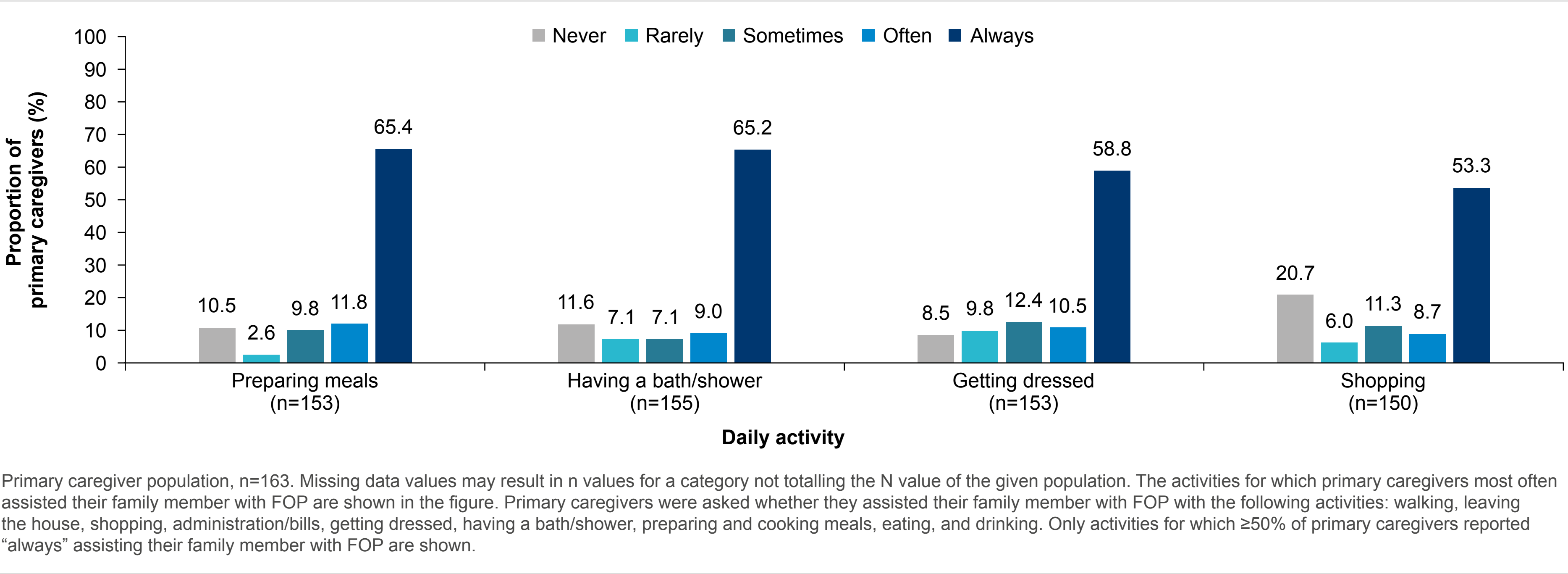
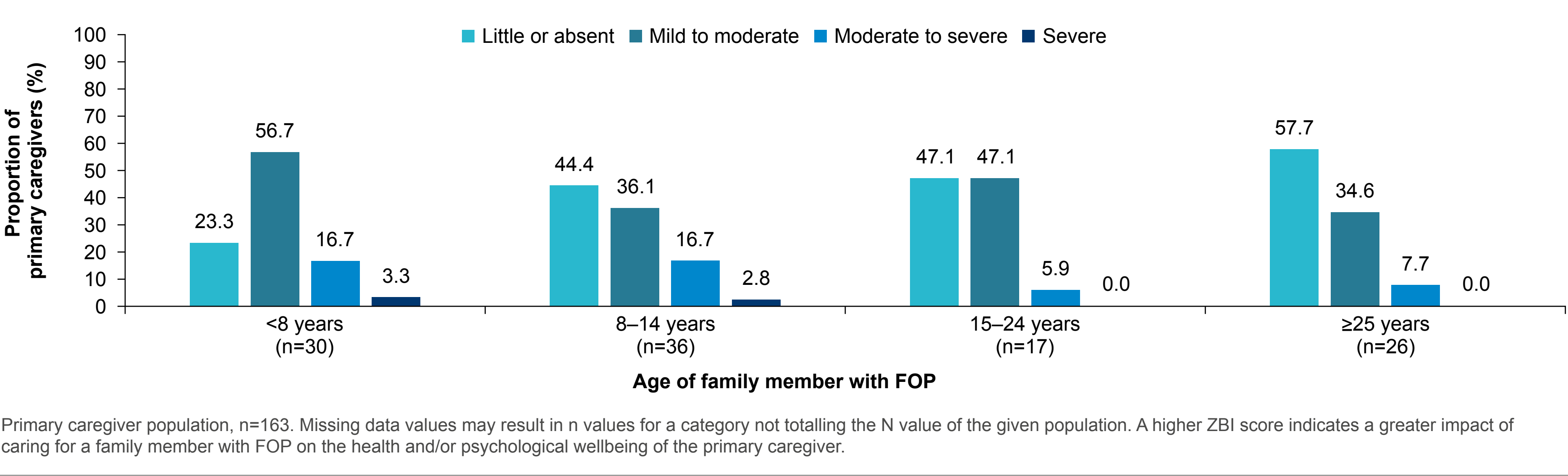


Figure 4. Impact on primary caregivers' health and/or psychological wellbeing (ZBI total score) by age of the family member with FOP



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