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XLH Australia

Understanding the
experiences of people
with XLH

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Introduction

In December 2021 and January 2022, XLH Australia (in collaboration with Kyowa Kirin Australia) conducted a nationwide survey of patients and carers of those living with X-linked hypophosphataemia (XLH).

The aim of the survey was to increase understanding of what it is like to live with XLH.

A quick **overview** of this report...

The survey results are broken down into three parts...

Living with XLH

- Diagnosis
- Impact of physical symptoms
- Attitudes
- Mental health impact
- Hopes for the future

XLH Treatment

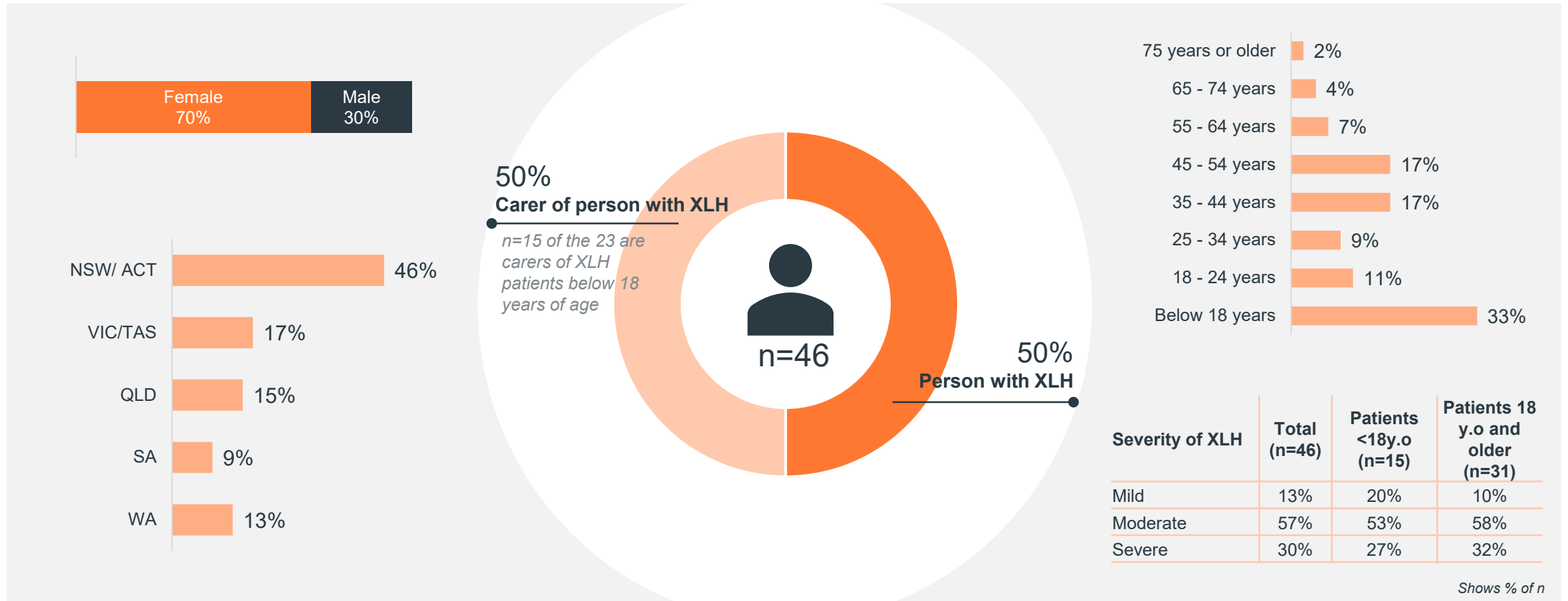
- Cost of living with XLH
- New treatment awareness
- Attitudes towards new treatments

The Surgery Experience

- Total surgeries undergone
- Osteotomies undergone
- Major dental surgeries

A total of n=46 people participated, with a straight split of carers and people with XLH, and with majority experiencing moderate or severe XLH...

Demographic profile of our XLH participants

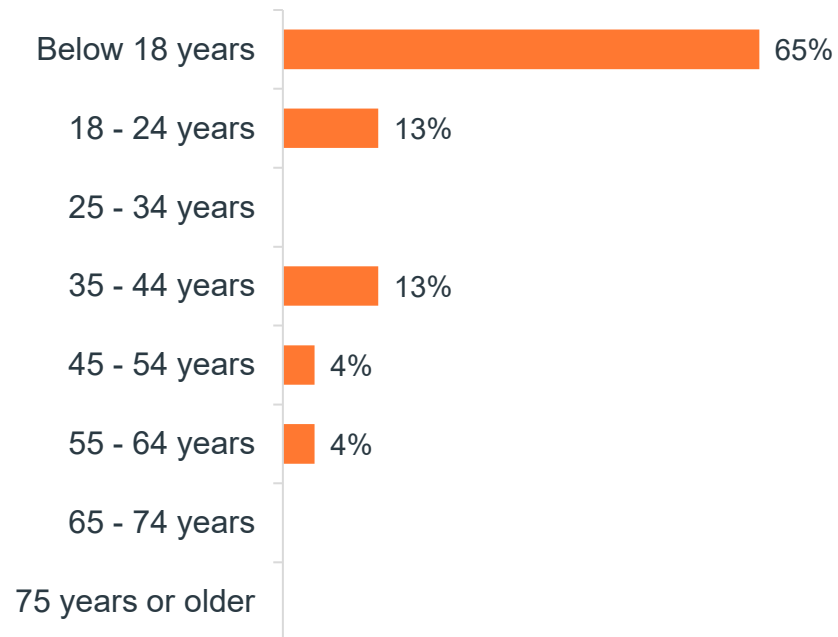


As expected, majority of the carers (65%) care for a person with XLH who is younger than 18 years of age

Carers of a person with XLH – Demographics of person with XLH they care for

(base: n=23)

Age of person with XLH they care for (base: n=23)



Severity of person with XLH they care for

Severity of XLH	Total (n=46)	Carers of a person with XLH (n=23)	Patient with XLH (n=26)
Mild	13%	17%	9%
Moderate	57%	57%	57%
Severe	30%	26%	35%

Key headlines...

XLH patients are diagnosed young and living with it is difficult for many

Majority of patients feel burdened by the physical and mental impacts of XLH, and most have had surgeries relating to XLH

Osteotomies and dental surgeries are common for patients to undergo. Osteotomies have long recovery period (more than 6 months for about a third)

Lack of awareness is believed to be impacting support, health funding, and understanding of the condition overall

Positively, most patients are currently on treatment and are aware and keen about new treatments

There is opportunity to raise awareness for XLH to encourage support from the populace, help create 'noise' for health funding, and to help boost patients' morale overall

Living with XLH

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On average, people with XLH were diagnosed at **3 years old**, and primarily diagnosed by a **endocrinologist**...

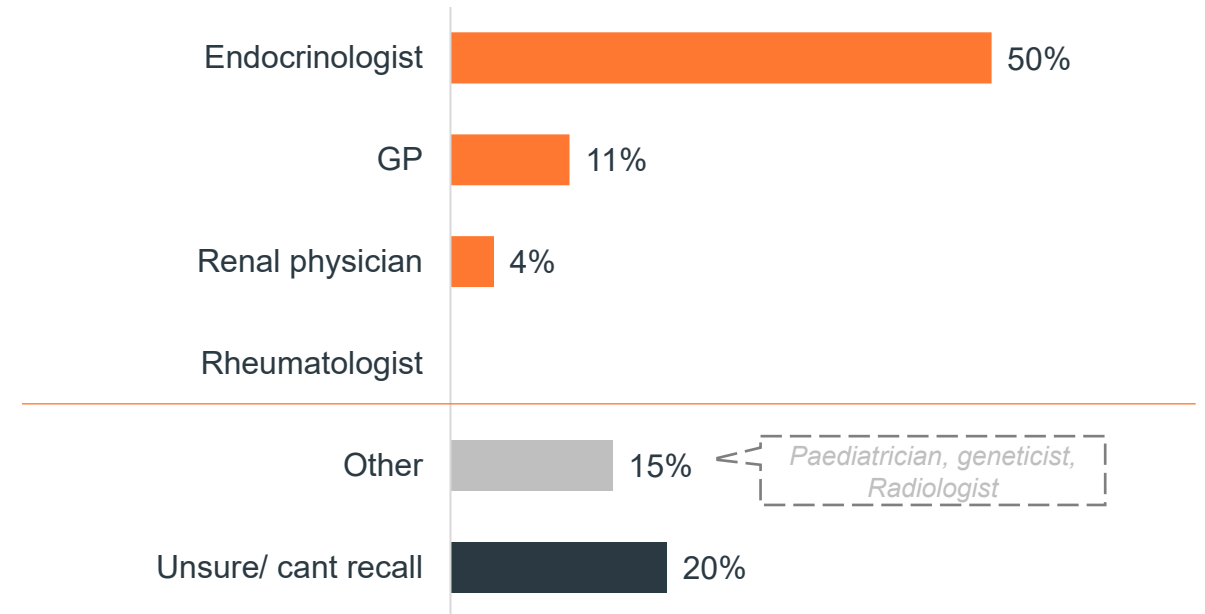
The Diagnosis

Age at diagnosis



Range: Less than 1 year old – 23 years old
Median: 2 years old
Mode: 2 years old

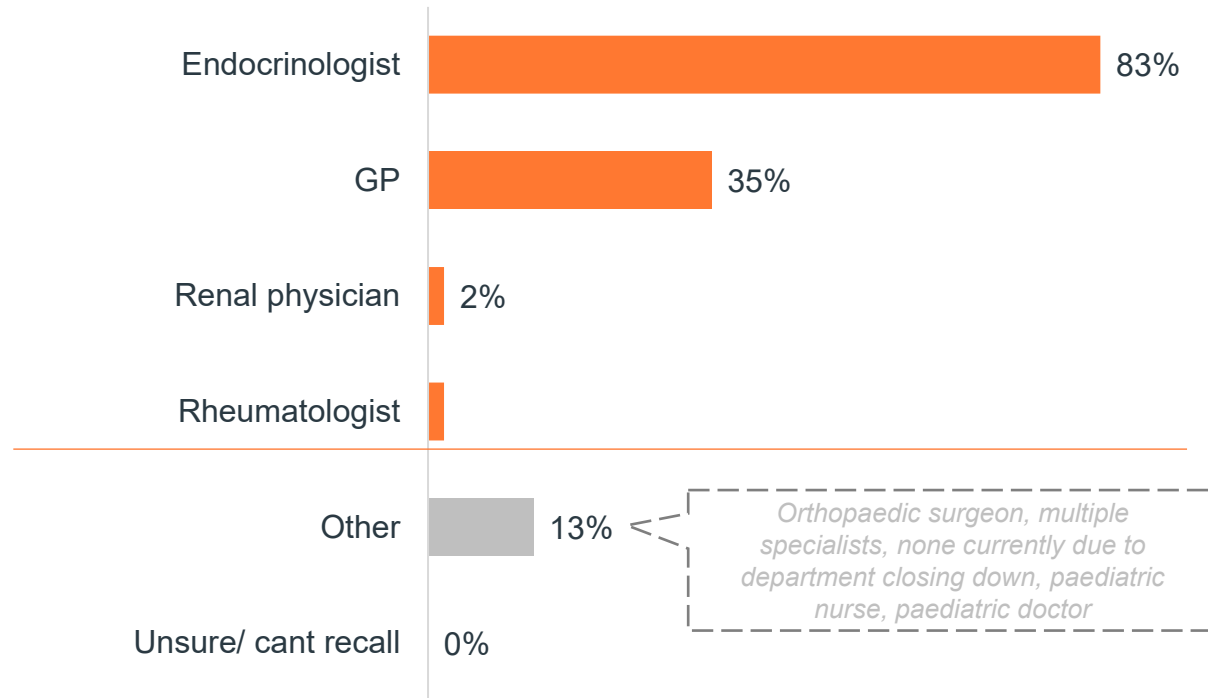
Source of diagnosis



Shows % of n

8 in 10 people with XLH are currently being managed by an endocrinologist...

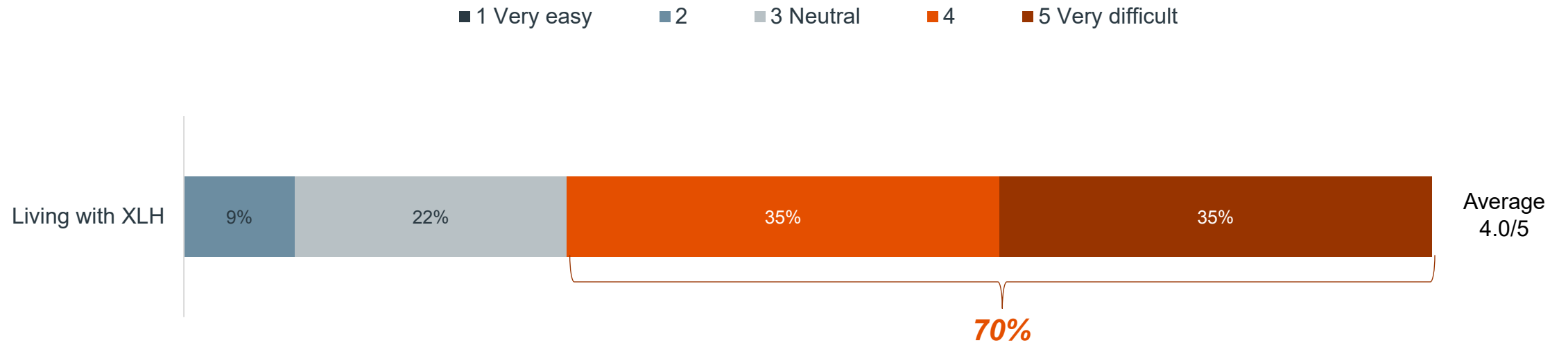
Current specialist



Shows % of n

7 in 10 people with XLH feel that the condition is difficult to live with

Living with XLH – Overall Attitude

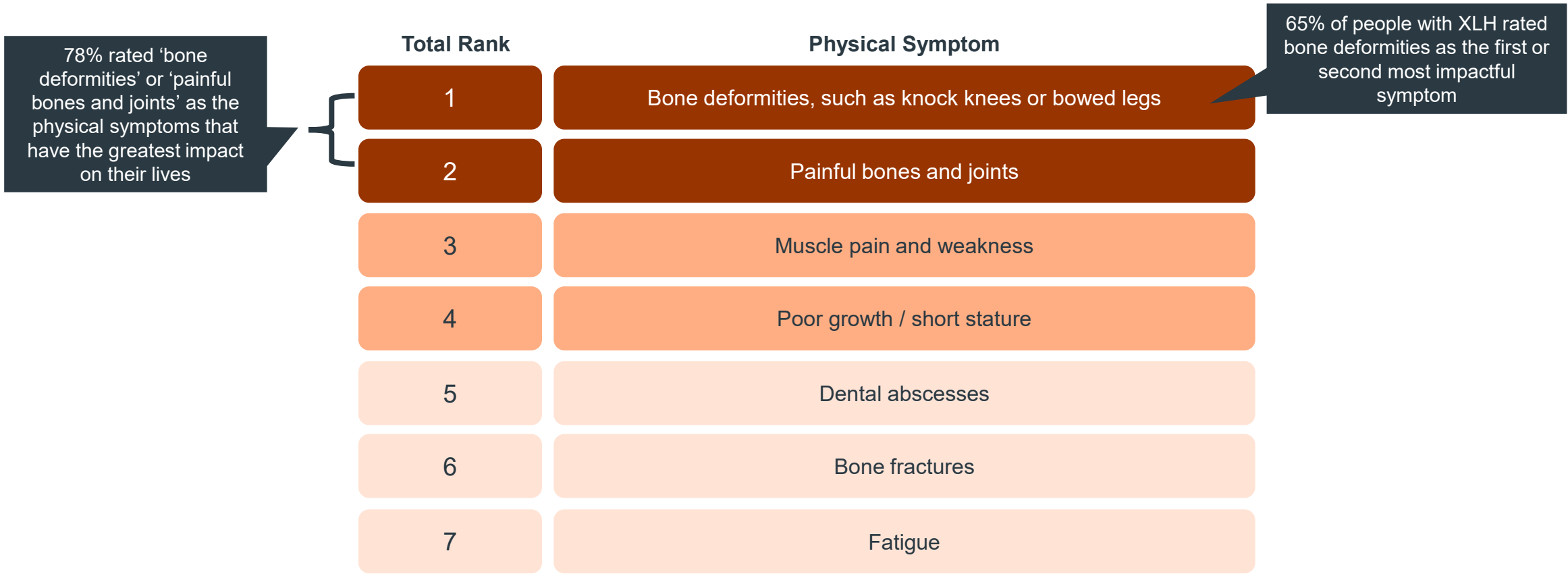


- Difficulty increases with severity, with severe having an average of 4.6/5 vs. those with mild having an average of 3.2/5 and moderate having 3.8/5
- Age does not really impact difficulty with no significant differences between those who are under 18 y.o and 18 y.o or older

Shows % of n

Bone deformities and painful bones/joints are the top two physical symptoms that impact people with XLH the most...

Living with XLH – Impact of physical symptoms

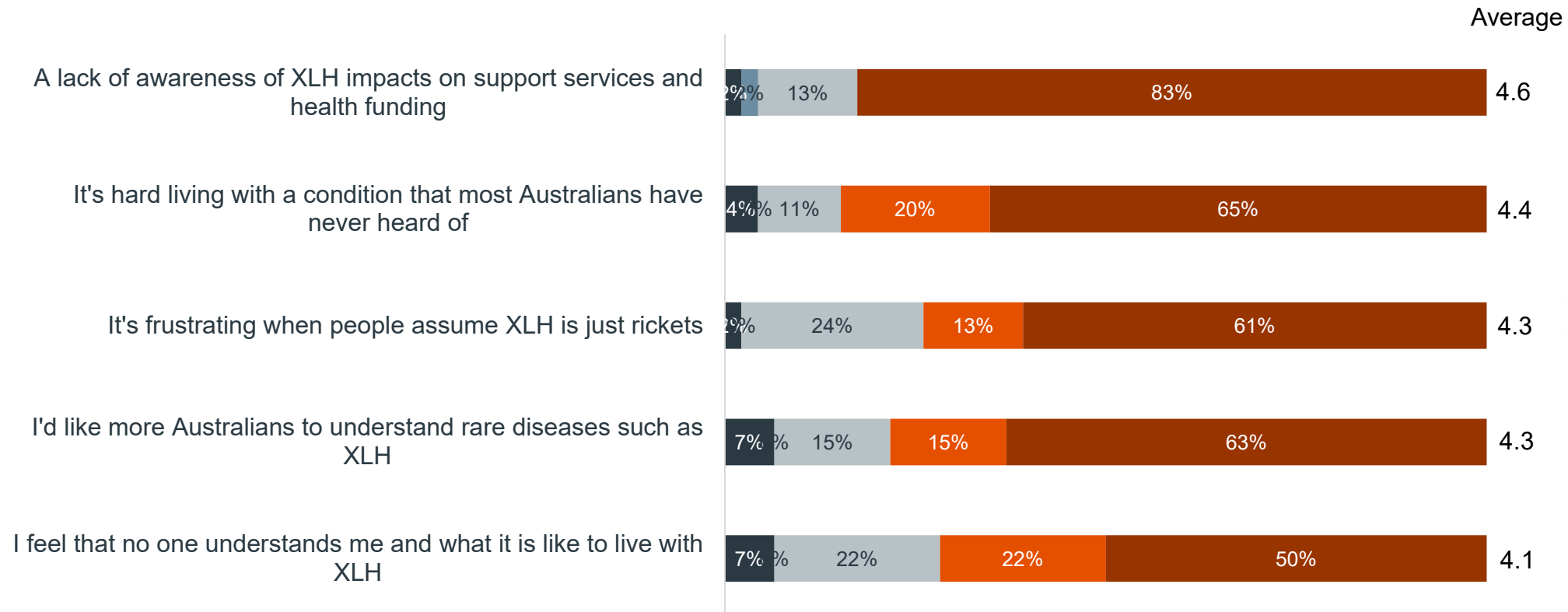


- Very similar ranking between younger than 18 y.o and 18 y.o or older

8 in 10 people with XLH feel the lack of awareness around XLH directly impacts the support services and funding available to them....

Living with XLH - Attitudes

■ 1 Strongly disagree ■ 2 ■ 3 ■ 4 ■ 5 Strongly agree



Lack of awareness is clearly an issue that affects people with XLH both tangibly and mentally

Shows % of n

E.

Source: Q4. Please indicate the extent to which [P: you agree or disagree / C: the person you care for agrees or disagrees] with the following statements. Rate on a scale of 1 to 5 where 1 = strongly disagree and 5 = strongly agree.

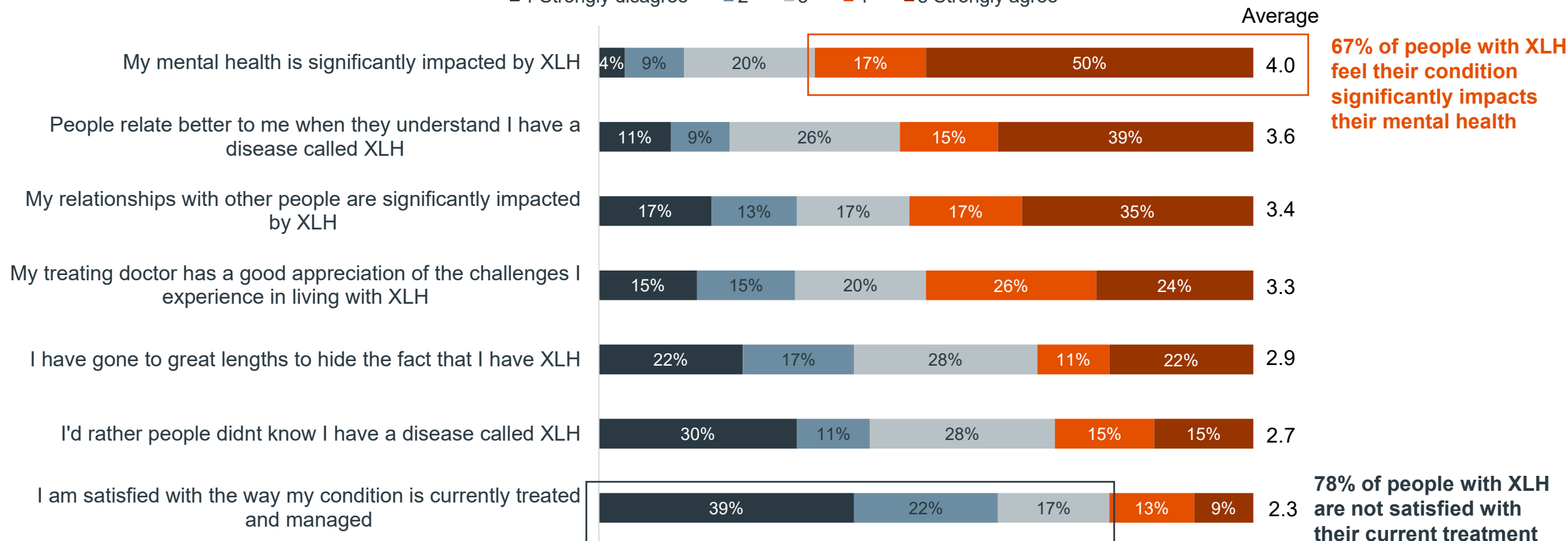
Base: n=46

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Almost 7 in 10 people with XLH feel their mental health is significantly impacted by XLH...

Living with XLH - Attitudes

■ 1 Strongly disagree ■ 2 ■ 3 ■ 4 ■ 5 Strongly agree



Shows % of n

E.

Source: Q4. Please indicate the extent to which [P: you agree or disagree / C: the person you care for agrees or disagrees] with the following statements. Rate on a scale of 1 to 5 where 1 = strongly disagree and 5 = strongly agree.

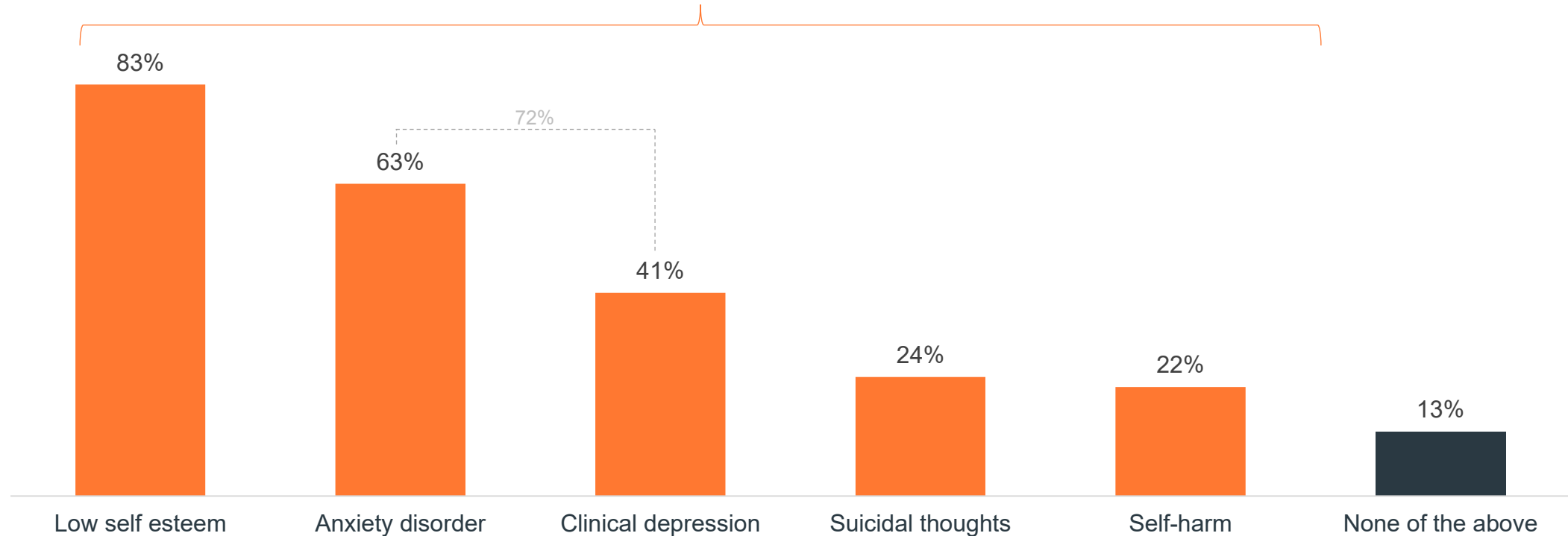
Base: n=46

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Low self esteem is the most common mental health issue experienced by people with XLH, but almost 9 in 10 have experienced a mental health issue of some kind at some stage...

Living with XLH – Mental health impact

87% of people with XLH have experienced a mental health issue at some stage



Comparative statistics: 1 in 5 (20%, or 4.8 million) Australians reported that they had a mental or behavioural condition during the collection period (July 2017 to June 2018) [AIHW National Health Survey 2017-18]

Shows % of n

E.

Q7. Thinking further about the impact of XLH, which of the following have [P: you / C: the person you care for] experienced at some stage during your life?

MULTI CHOICE Select all that apply

Base: n=46

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More than half of people with XLH find the mental and physical impacts of XLH are equally burdensome to deal with...



54%

The physical and emotional impacts are equally burdensome

33%

The pain, discomfort and restricted mobility are the greatest challenges

11%

The emotional and mental burden are the greatest challenges

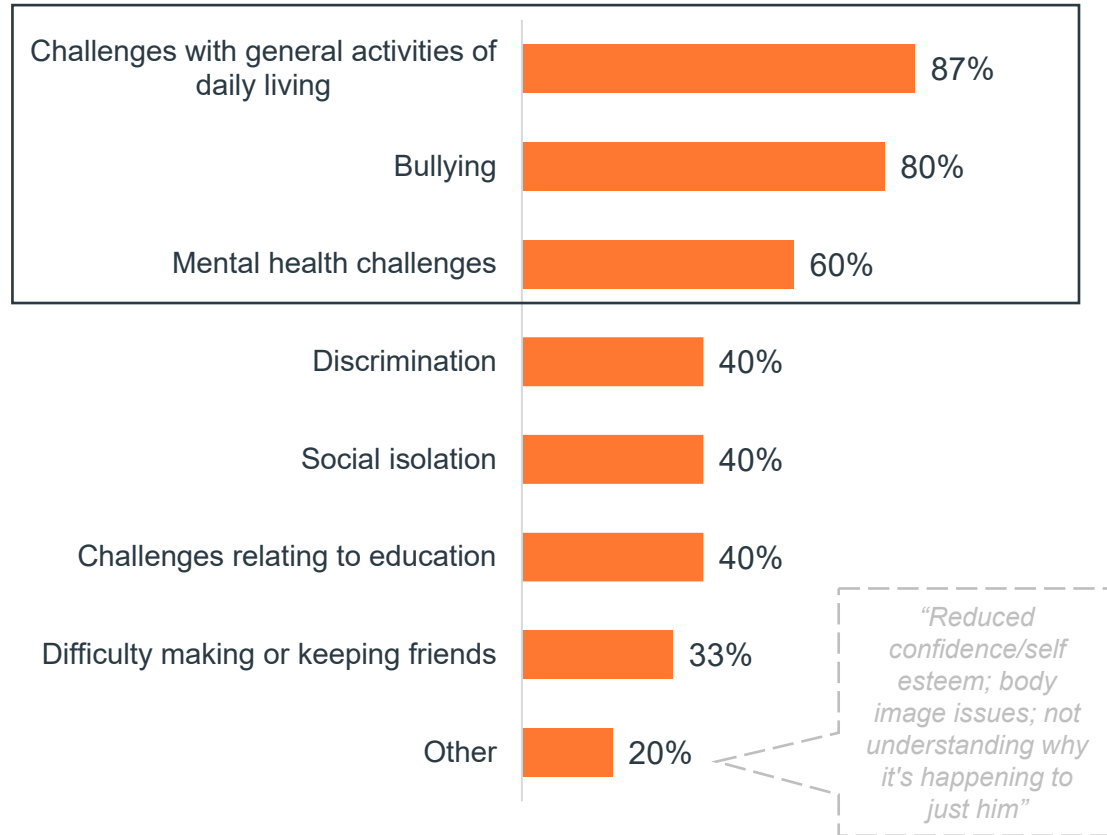
2% said none of the above

Shows % of n

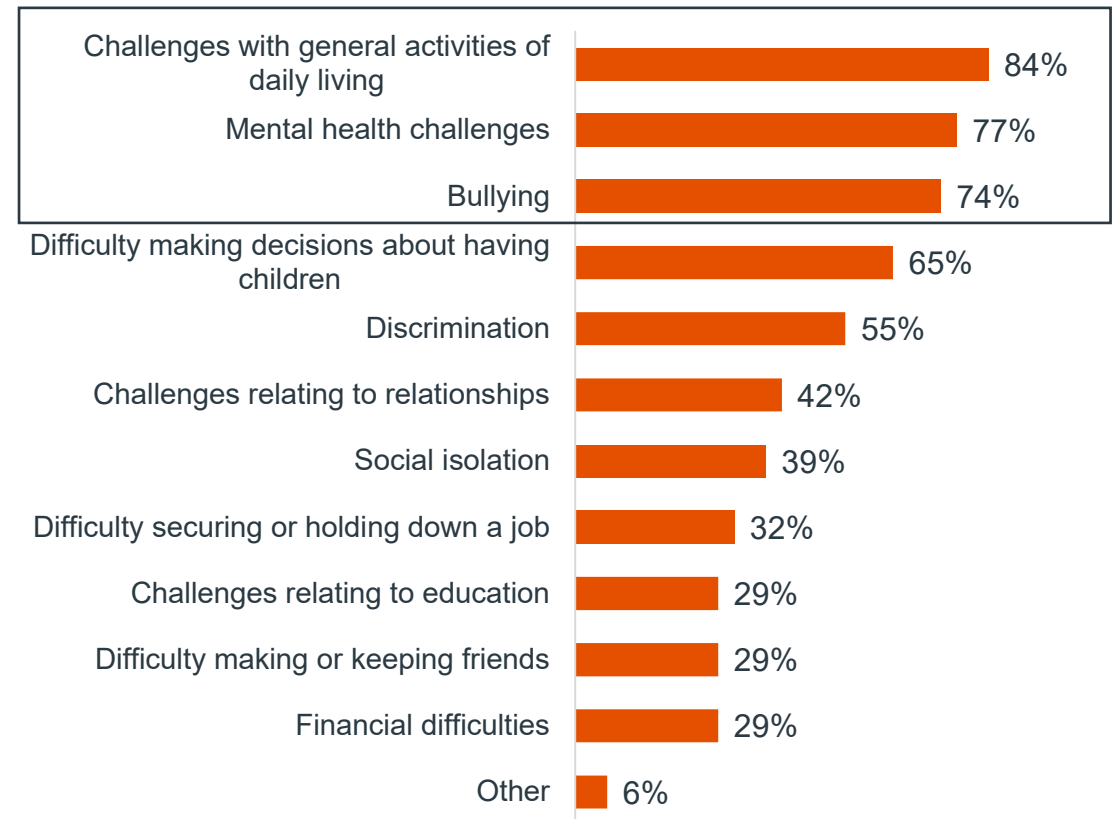
Almost all experience challenges with every day living and concerningly, mental health challenges and bullying are top ranked regardless of age

Living with XLH – Challenges experienced as a result of XLH

Under 18 y.o



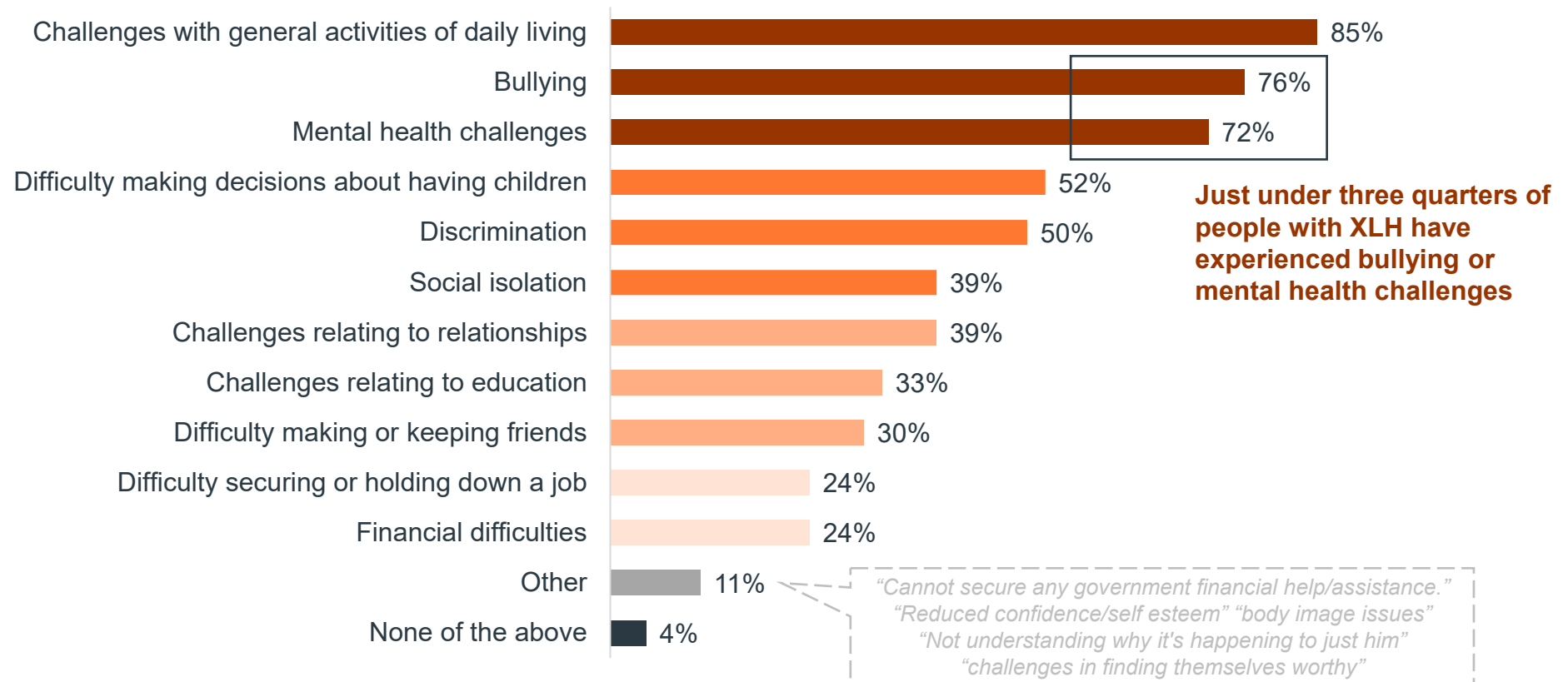
18 y.o and over



Shows % of n

Almost all experience challenges with every day living but concerningly, **just under three quarters** of people with XLH have experienced bullying or mental health challenges...

Living with XLH – Challenges experienced as a result of XLH



Shows % of n

E.

Source: Q6. Please indicate which of the following [P: have you experienced / C: has the person you care for experienced] as a result of living with XLH?
 Select all that apply.

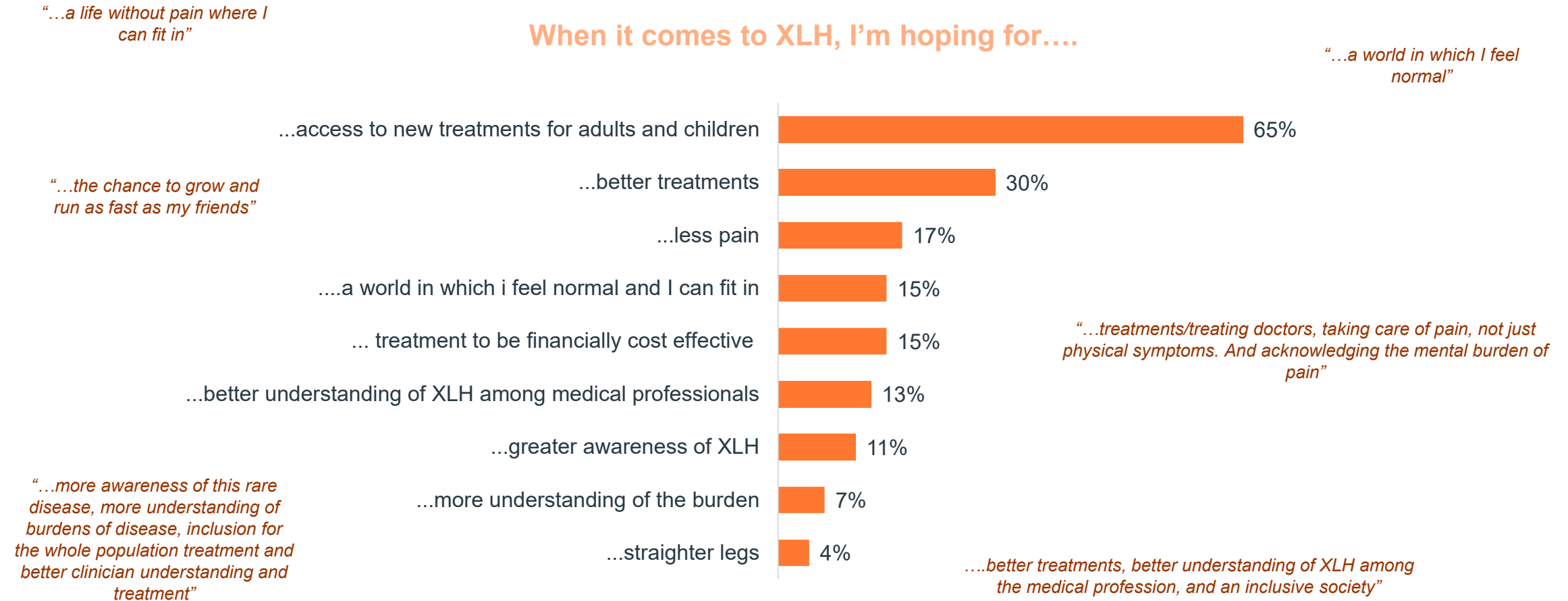
Base: n=46

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Looking to the future, **two thirds** of people with XLH are hoping for access to new treatments for both adults and children...

Living with XLH - Hopes for the future

When it comes to XLH, I'm hoping for....



Shows % of n

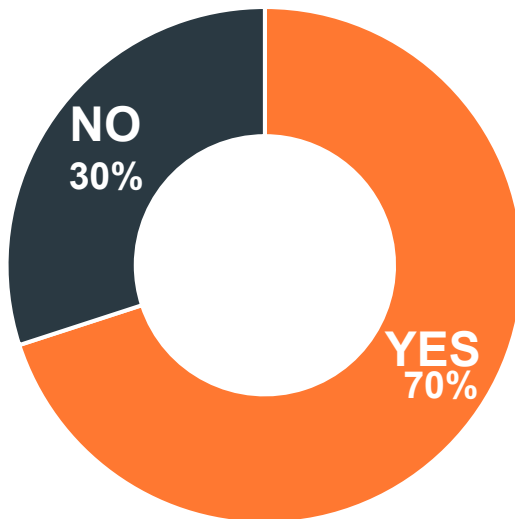
Treatment for XLH

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Three quarters of people with XLH are currently on treatment, but are spending varying amounts on XLH in an average year....

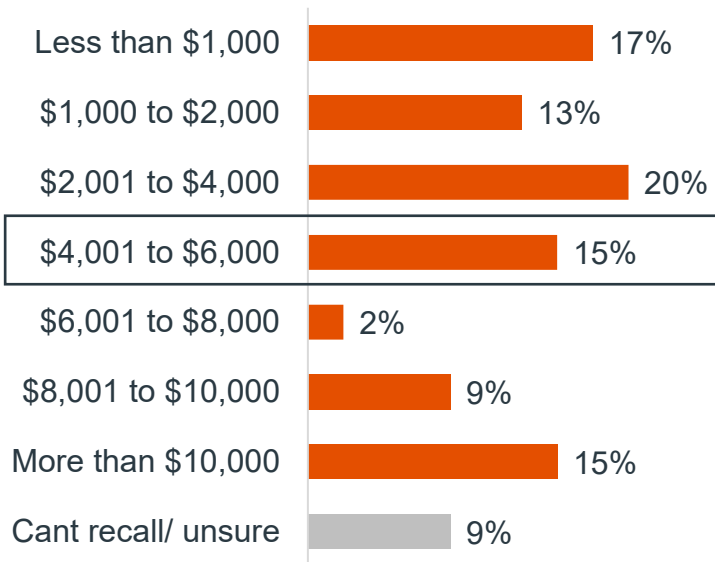
Current treatment

Currently on treatment



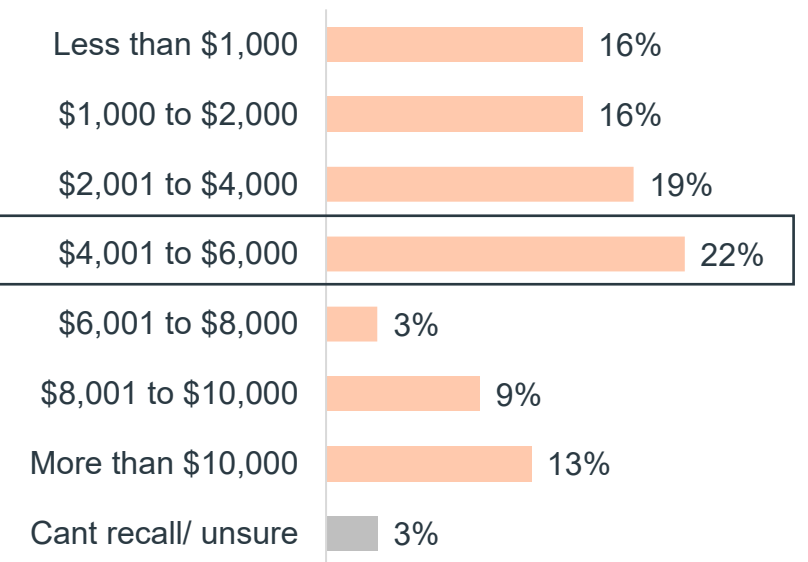
Average cost of living with XLH in an average year - total

Including medical appointments, treatments, equipment and services



Average cost of living with XLH in an average year – those on treatment (n=32)

Including medical appointments, treatments, equipment, services and medication



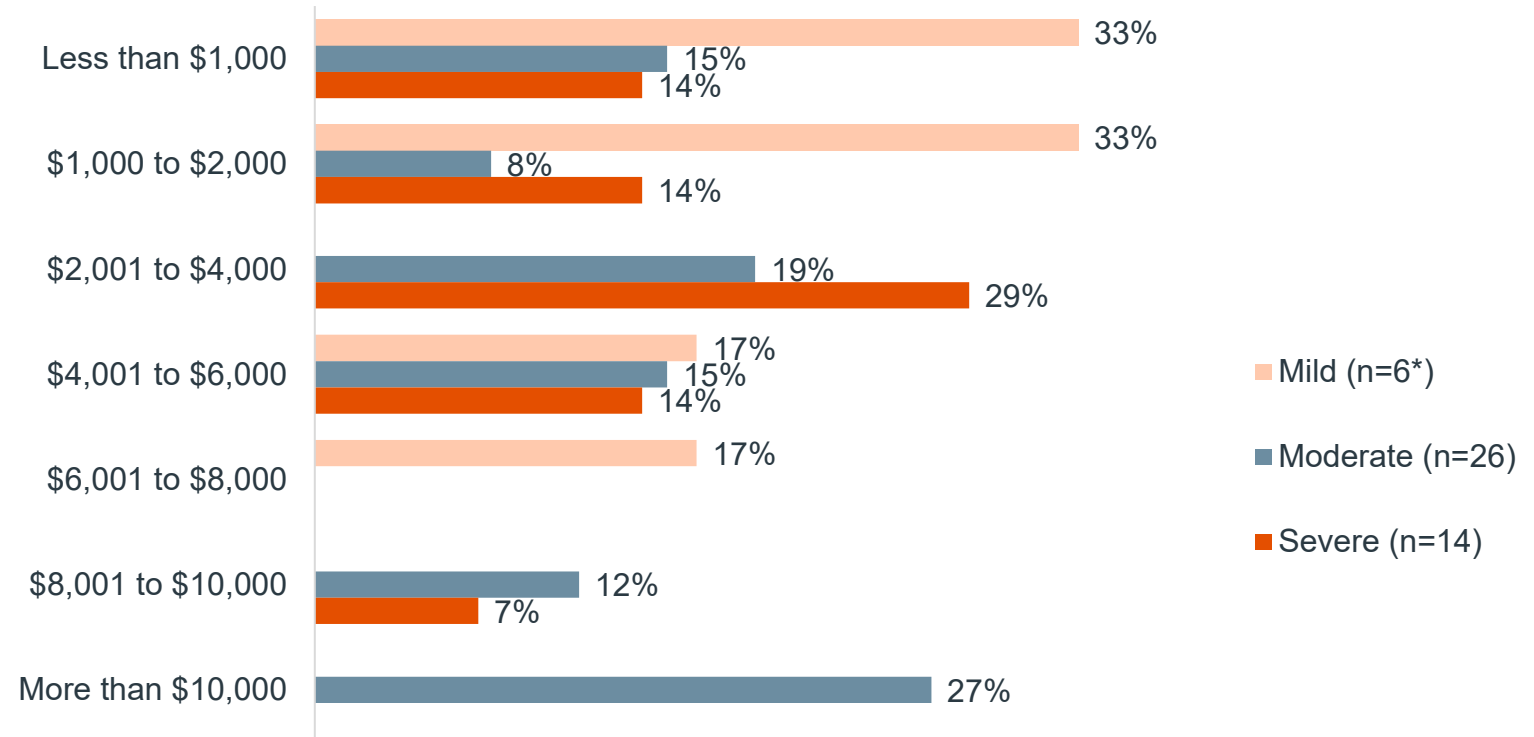
Estimated average cost of \$5,309 per year

Approximate costs also vary by severity – more costly for those with a severe condition

Average cost by severity

Average cost of living with XLH in an average year

Including medical appointments, treatments, equipment and services



E.

Q8: Now thinking about the cost involved in living with XLH, approximately how much [P: do you spend / C: is spent on the person you care for] in an average year on medical appointments, treatments, equipment, and services relating to XLH? Q9: Are [P: you / C: the person you care for] currently receiving treatment for XLH?

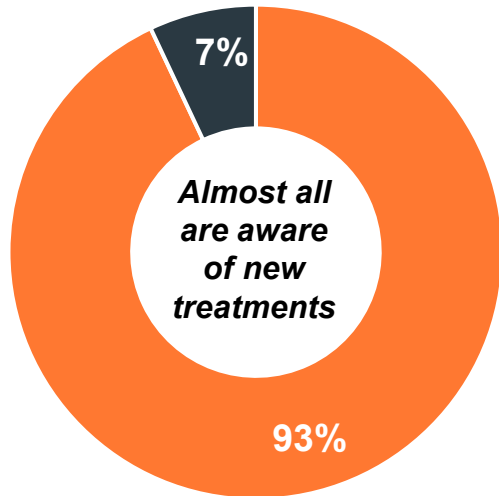
Base: n=46

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9 in 10 people with XLH would like to know more about new treatments and are willing to try these new treatments if their doctor recommends it

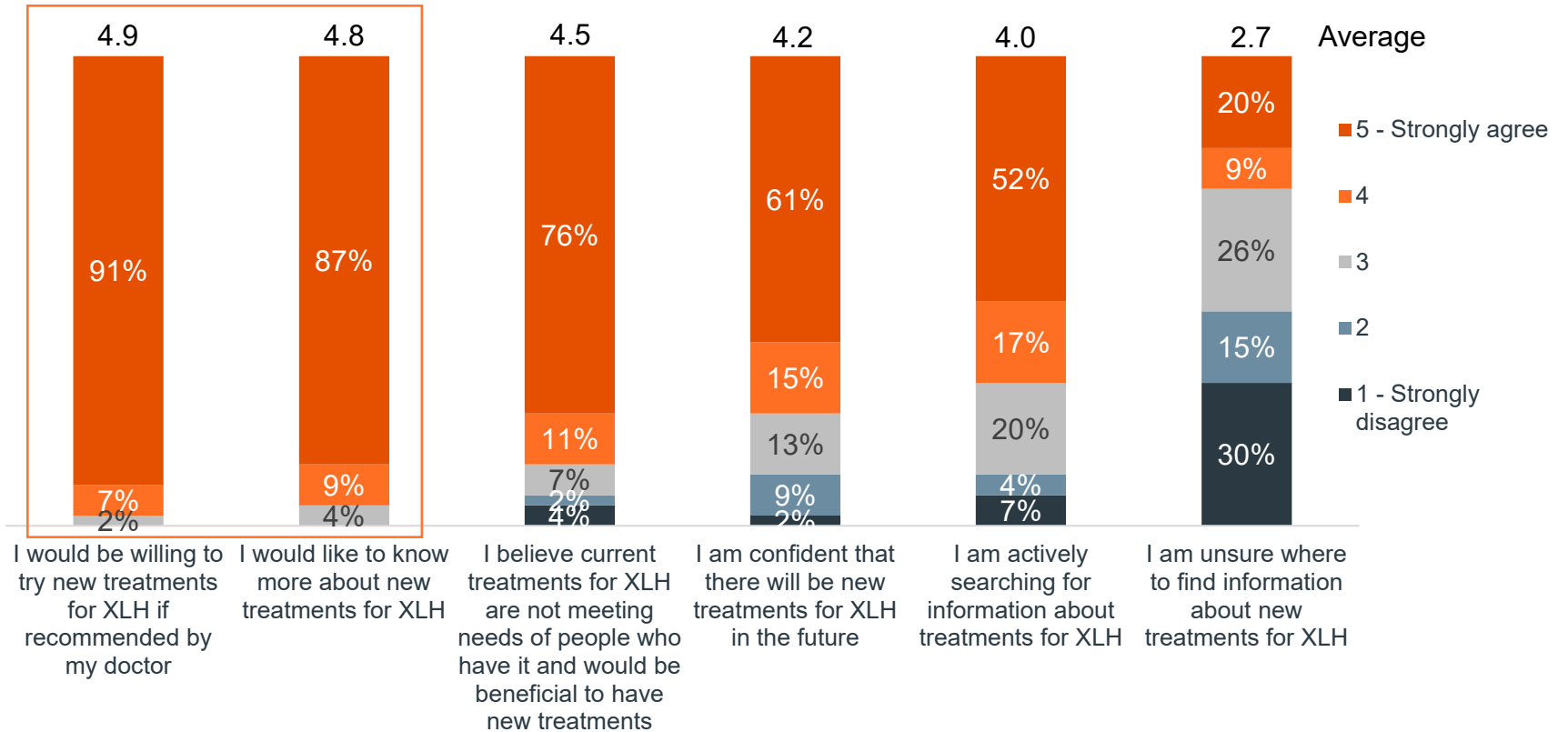
New treatments

Awareness of new treatments



Almost all want to know more and are willing to trial new treatments

Attitudes towards new treatments



E.

Q10: Are [P: you / C: the person you care for] aware of any new treatments for XLH? Q11: Please indicate below to what extent do [P: you / C: the person you care for] agree with the following statements. Rate on a scale of 1 to 5 where 1=strongly disagree and 5=strongly agree.

Base: n=46

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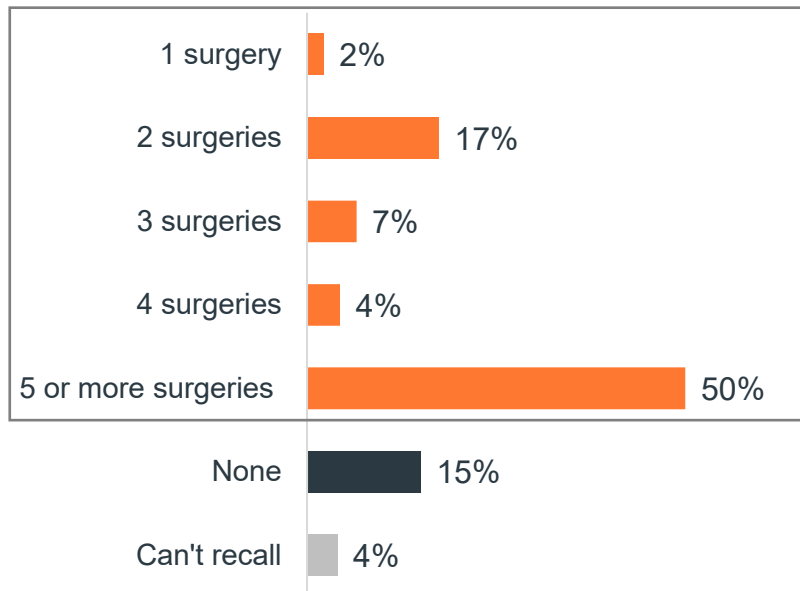
The surgery experience

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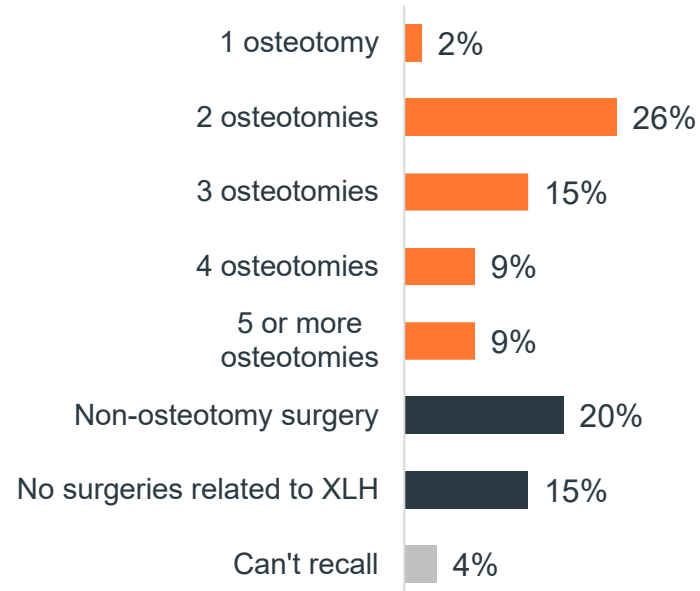
More than half of people with XLH have undergone 5 or more surgeries relating to XLH, with these mostly constituting 5 or more dental surgeries and 2 osteotomies

Surgeries undergone

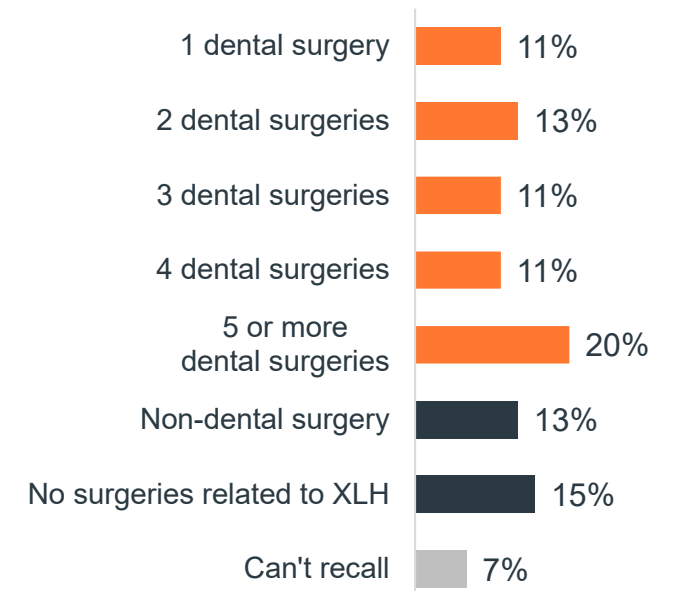
Number of surgical procedures relating to XLH undergone (total)



Number of osteotomies undergone (total)

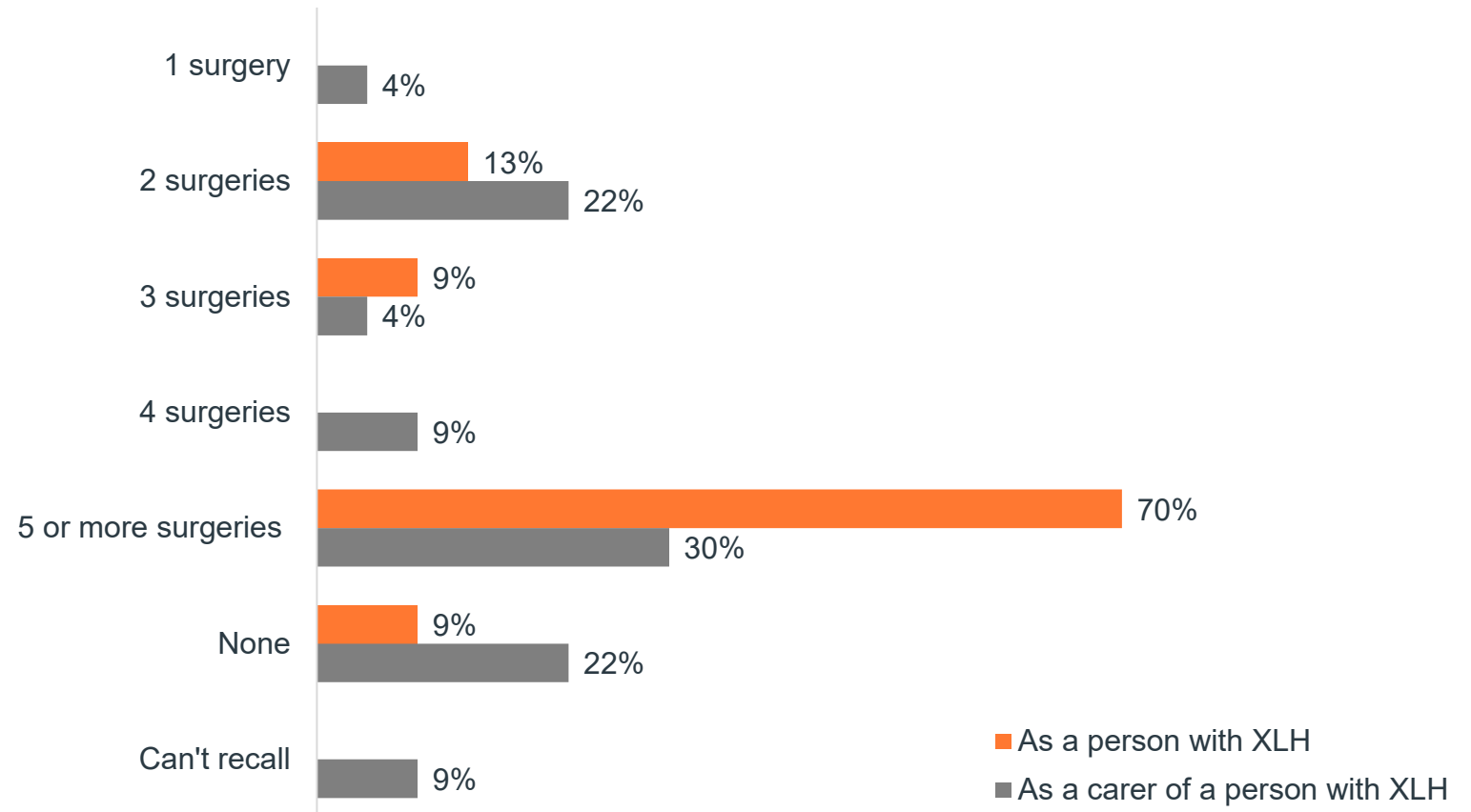


Number of major dental surgeries (total)



A greater percentage of people with XLH reported undergoing 5 or more surgeries related to XLH versus carers of a person with XLH...

Surgeries related to XLH undergone – Carers vs Person with XLH



On average, people with XLH have their first osteotomy during their teen years and their last in their early 20's. It can take them **3 months or longer** to walk after the surgery

Osteotomies

Base: n=28 who have undergone a osteotomy

Age of first and last osteotomy performed

14 years

average age when FIRST osteotomy occurred

Range: 4 years old to 37 years old

Median: 14 years

Mode: 5 years old

23 years

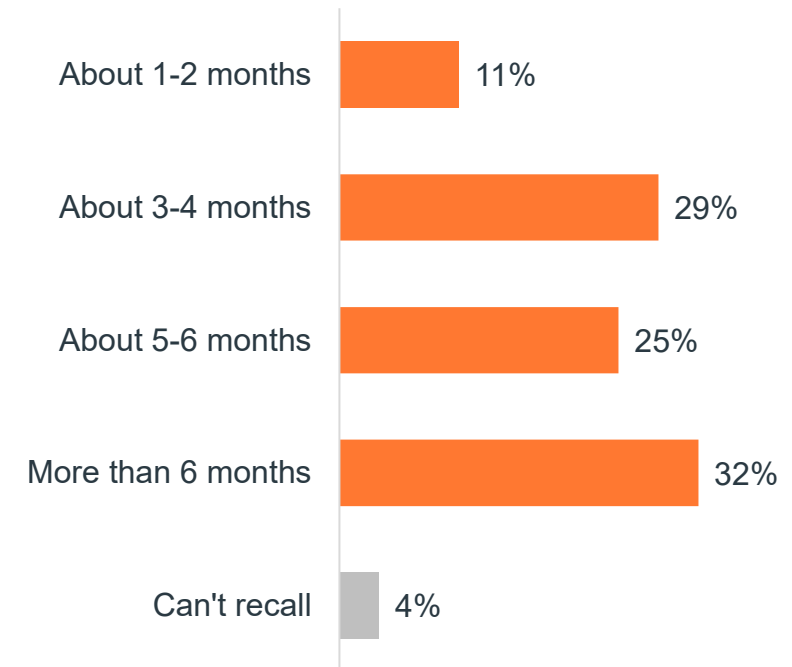
average age when LAST osteotomy occurred

Range: 7 years old to 58 years old

Median: 19 years old

Mode: 19 years old

Length of time to recover and walk unassisted after osteotomy

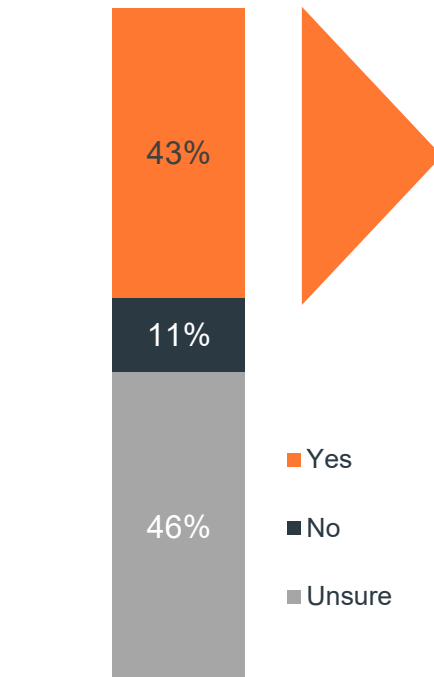


Just under half believe they will require a osteotomy in the future, but most are unsure when that next osteotomy will be...

Future Osteotomies

Base: n=28 who have undergone a osteotomy

Likelihood of future osteotomies



Date of next osteotomy (verbatim)

"March 2022"

"2022" (n=2)

"Not sure - I tried to have it done on my femur, but once the surgeon got in there to do it, he decided it was too risky and aborted the procedure"

"Re assess in 12 months"

"2 years"

N=6 unsure of when their next osteotomy will be

On average, people with XLH are in their **teens** when they had their first dental procedure and are in their **30's** for their last...

Dental Surgery

Base: n=30 who have undergone a major dental surgery

Age when dental procedure occurred

15 years

average age when FIRST major dental procedure occurred

Range: 4 years old to 39 years old

Median: 10 years old

Mode: 5 years old

30 years

average age when LAST tooth removal occurred

Range: 4 years old to 69 years old

Median: 26 years old

Mode: 19 years old

Number of teeth removed (Base: n=25)

4 baby teeth

on average have been removed

Range: 0 to 20 teeth

40% haven't had any baby teeth removed

6 adult teeth

on average have been removed

Range: 0 to 30 adult teeth

28% haven't had any adult teeth removed

E.

Q17: How old [P: were you / C: the person you care for] when these major dental procedures first occurred (i.e. removal of teeth)?

Q17a: If [P: you have / C: the person you care for has] had multiple major dental procedures, such as tooth removal, how old [P: were you / C: was the person you care for] when the final tooth removal was performed?

Q17b: In total, how many teeth [P: have you / C: has the person you care for] had removed?.

Base: n=30 who have undergone a major dental surgery

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Summary

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The impact of XLH...

Overall, XLH is a difficult condition to live with.

People affected by XLH struggle with bone deformities and joint/bone pain, with half experiencing 5 or more surgeries relating to their XLH. Majority of those who underwent an osteotomy couldn't walk unassisted for 3 months or longer after the surgery.

A lack of awareness around the condition exacerbates their struggles, with 8 in 10 believing the lack of awareness is directly impacting support services available to them. Consequently, almost 9 in 10 people with XLH have experienced a mental health issue at some stage, with experiences of bullying being a highly prominent issue.

Looking to the future, individuals and carers of people affected by XLH are hoping for better access to new treatments for both adults and children, so they can live a normal, pain free life.

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Thank you.

ELBOW.

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A proud partner of
Rare Voices Australia

