1000 Voices

The impact of ageing on the lives of people with spina bifida

A report for Health and Social Care Professionals, and Commissioners

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Authors: Gill Yaz, Health Development Manager, Shine
Dr Jenny Smith-Wymant, Health Engagement Officer, Shine

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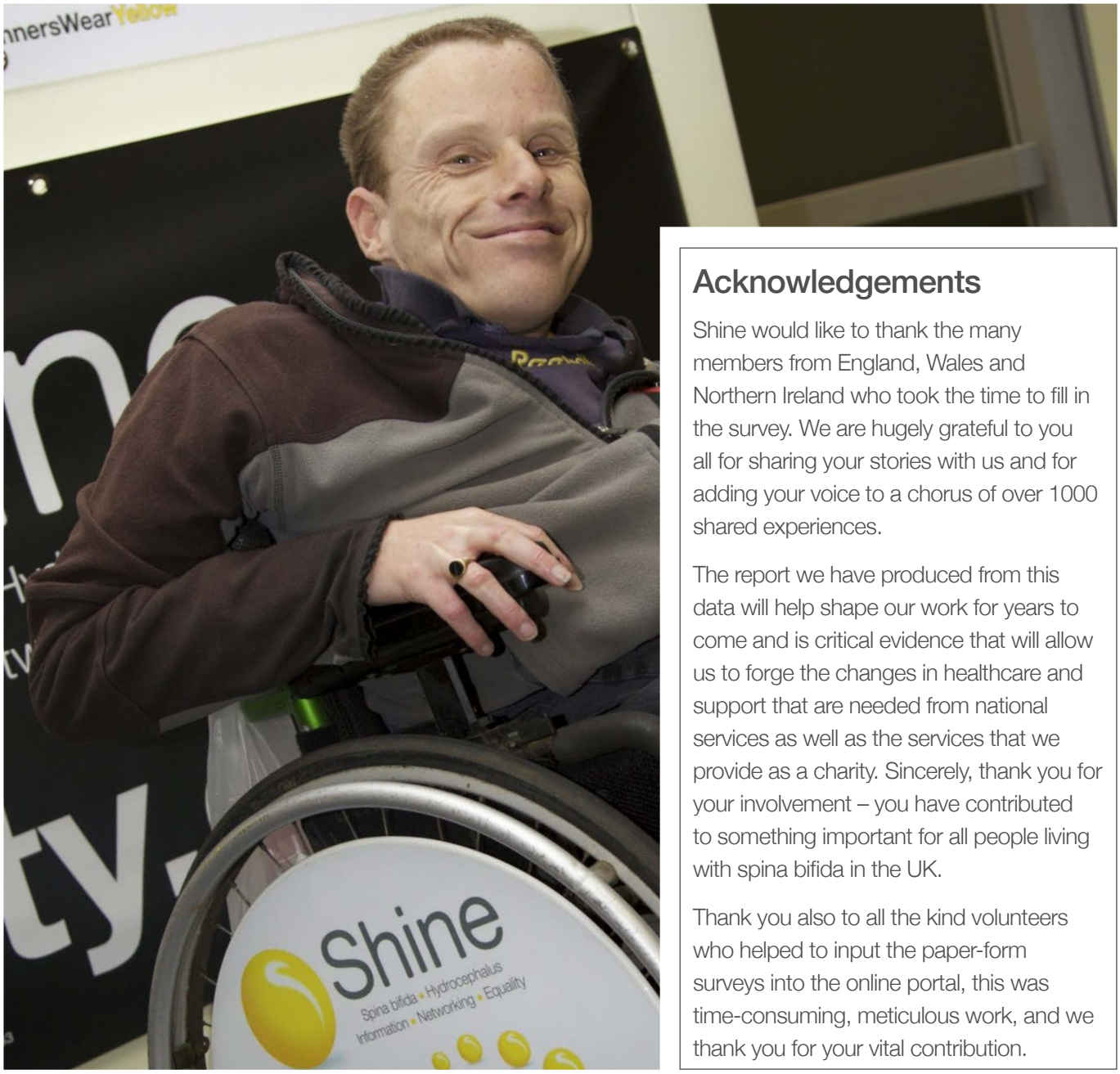
Founded in 1966, Shine is a national charity, working across England, Wales and Northern Ireland. We provide specialist support pre-birth and beyond for anyone living with spina bifida, hydrocephalus and other conditions such as anencephaly and encephalocele. Shine also advises and supports parents, families, carers, and professionals, notably across the health, care, welfare and education sectors.

Shine has a membership of over 12,000 individuals living with spina bifida and/or hydrocephalus. Individuals and their families can contact us for information and support at any time. We also work closely with professionals from health and social care who refer individuals or parents to Shine when they need us.

For further information about the work we do or to make a referral to Shine, or to discuss the findings of our research, contact us on 01733 555988 or email firstcontact@shinecharity.org.uk.



Acknowledgements

Shine would like to thank the many members from England, Wales and Northern Ireland who took the time to fill in the survey. We are hugely grateful to you all for sharing your stories with us and for adding your voice to a chorus of over 1000 shared experiences.

The report we have produced from this data will help shape our work for years to come and is critical evidence that will allow us to forge the changes in healthcare and support that are needed from national services as well as the services that we provide as a charity. Sincerely, thank you for your involvement – you have contributed to something important for all people living with spina bifida in the UK.

Thank you also to all the kind volunteers who helped to input the paper-form surveys into the online portal, this was time-consuming, meticulous work, and we thank you for your vital contribution.

Foreword by Roger Bayston

I am honoured to have been asked to write the Foreword for this report into the impact of ageing on the lives of people with spina bifida.

I began my career journey in the late 1960s when working with extremely eminent figures involved in spina bifida and hydrocephalus, such as professors John Emery and Robert Zachary. In the mid-1970s I was fortunate to be awarded a Research Fellowship by ASBAH (now Shine Charity) and this launched me into a career of research into problems of spina bifida and hydrocephalus. I have therefore been working with people with spina bifida for about 60 years. I have seen enormous changes in treatments and management of the condition, where the spine fails to form properly during development of the fetus. This leads to an array of problems with mobility, dexterity, and function of bladder and bowel, often compounded by accompanying hydrocephalus. The changes have been in response to new knowledge about the effects of earlier treatments. One difference is that there is now much more acceptance by clinicians that people with spina bifida who are able to do so might choose not to walk, especially as they approach teens, and elect for a wheelchair (in which they often develop amazing skill).

However, most of the older people with spina bifida underwent earlier treatments based on original conceptions and they are now growing old alongside all of us. There is now a great deal of interest in ageing, and for those with spina bifida there is concern that they might age more rapidly or differently due to their physical problems, and possibly due to hydrocephalus. For those with the condition, it is obviously vital that they are able to look ahead and plan for any enhanced needs, and for Shine Charity it is equally important that they can see ahead to how they might address these requirements. This is the reason for this survey of just over 1000 people with spina bifida, to find out how they manage mobility, medical issues, employment and social and personal life.

The survey was carried out in England, Wales and Northern Ireland. The data are presented in a very clear way, with charts and graphs but also with free comment from responders which is often extremely revealing beyond what the statistical data show.

A decline in mobility was reported by 78% of responders as they aged, but the survey showed that much of the change occurred around the age of 35yrs. While some enjoyed using a wheelchair, many regretted their loss of ability to walk. Musculoskeletal pain was a limiting factor in many cases, and if this could be reduced or delayed, it might make a considerable difference to quality of life, making the case for more attention to lifestyle-integrated exercise.

Skin problems such as pressure ulcers are a little-recognised problem but were highlighted by the survey. Sadly 23% of responders reported that they had acquired a pressure sore while in hospital for another reason.

Health services for people with spina bifida need to be improved so that they receive specialist attention where needed. Even 40 years ago, children with spina bifida could expect regular contact with specialist paediatricians, urologist and neurosurgeons working as a team, but on attaining the age of 16yrs they have to join adult care which is fragmented so that they lose the attention of a dedicated team of specialists. Now such combined specialist care for children is uncommon.

The authors are very well qualified to produce this report:

Gill Yaz is Health Development Manager at Shine Charity and has been with the charity for 26 years. She is a registered nurse with a special interest in continence problems, but her experience and knowledge extend far beyond this. Gill has been active in hydrocephalus and spina bifida research for many years and has served on the Executive Committee of an international research society. She is also a member of the Unplanned Admissions Consensus Committee, devoted to improvement in continence care and dissemination of best practice. She also serves on National Institutes of Health Trial Monitoring Committee in connection with a study of urinary tract infection in catheter users.

Dr Jenny Smith-Wymant is Health Engagement Officer at the charity. She has a PhD in cell and molecular biology, a background in research engagement, and is well equipped to bring stringent scientific input to Shine and to this survey. Jenny produces educational materials and is involved in fostering stakeholder networks for Shine Charity.

This report is aimed at professionals. Sadly, there is still a lack of knowledge and understanding among health professionals of all aspects of spina bifida, and it does not feature strongly, if at all, on medical school curricula. The report is clear and readable, and it is hoped that all health professionals involved in care of those with spina bifida will read it and use the data to increase their understanding of the issues, and to improve the standard of care for those with the condition.

Roger Bayston

Professor of Surgical infection
Chief Investigator, NIHR CAUTI Trial
Co-Lead Investigator, NIHR CAP study
Head, Biomaterials – Related Infection Group
Division Research Impact Lead
School of Medicine
University of Nottingham UK
President, Nottingham Medico Chirurgical Society

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Introduction by Kate Steele, Shine CEO

Spina bifida is a developmental condition affecting the brain and spine, often leading to physical and cognitive impairments, and bladder and bowel issues. Widely regarded as one of the most severe conditions compatible with life, open spina bifida can result in significant morbidity, with numerous body systems and tissues affected. It was only from the late 1950s that babies born with spina bifida began to survive in significant numbers, as the invention of the shunt for hydrocephalus made treatment effective for the first time. In the early days, many young children succumbed to infections and renal failure, but as treatment improved, so did survival.

Dedicated teams of paediatric surgeons developed treatments and services to see the young people safely through to adult life, albeit with severe physical and cognitive impairment in many cases. However, there has never been the systematic, coordinated health care in adult services that developed so successfully in paediatrics. Many adults with spina bifida are now in their middle years, or beyond, and many have not had their condition monitored since leaving children's services. So, what happens to people with spina bifida in the intervening years, and what is the impact of years of fragmented and ad hoc healthcare? What are the implications for the healthcare services who have to pick up the pieces when adult patients present later in life with a myriad of complex issues exacerbated by inconsistent care?

In 2020, Shine surveyed its members with spina bifida. All adults over 25 were asked for their experiences of living with their condition, changes as they age, their health services, and how their needs are met by health services.

Over 1000 responses were received. The results show that, although young people may be discharged from regular monitoring by health professionals at age 18, their health often deteriorates, mobility declines, pain increases, and emergency admissions for pressure ulcers, sepsis or UTI increase. Although spina bifida is a non-progressive condition, the impact of tethered cord, Chiari II, use of mobility aids, and the sequelae of insensate skin and immobility mount up. Numerous adults reported that many aspects of their health, ability, and quality of life declined from around the age of 35.

Many stopped driving, others stopped working, anxiety and depression worsened, at an age many non-disabled adults regard as their prime.

The complex interplay of many aspects of the condition means that without a thorough, holistic, multidisciplinary approach, opportunities for preventing ill health and emergency hospital admissions will be missed. For example, a young adult with symptoms of tethered cord may experience pain, deterioration of bladder and bowel function, reduced mobility and sensation, foot deformity, pressure sores, cellulitis, lymphoedema and osteoporosis.

They may be under different healthcare professionals for each individual complaint, but with no real plan to join the dots. The care is largely reactive, waiting until a problem is sufficiently advanced to warrant referral. The huge drawback of this approach is that delays in addressing problems makes treatment more difficult and costly, and the health of the patient suffers.

In our survey some conditions, such as osteoporosis and obstructive sleep apnoea, while more common in people with spina bifida, still appeared greatly underdiagnosed. Adults in their 50s and older reported not having blood pressure or prostate checks. Opportunities to prevent ill health are being missed at every level of our healthcare system, with serious human and financial cost.

We can and must improve care for adults with spina bifida by planning proactive, integrated services from transition throughout adulthood. Shine asks you to consider how you can improve your services to do this. Please share this report with colleagues and ask them to do the same.

1.1 Background

Spina bifida is a condition caused by a difference in embryo development where the structure called the neural tube fails to close completely. The neural tube is the basic scaffold from which the spine and brain develop.

As the fetus matures, the spinal column forms with an open lesion and often results in changes to the way the central nervous system functions. Effects differ from person to person, but sensation, movement, bladder and bowel function, balance, and cognition are often affected to varying degrees. Survival to, and throughout, adulthood of people with spina bifida has improved greatly since the 1950s; the invention and wide use of the shunt has especially improved life expectancy. The relatively recent increase in survival with spina bifida means that we are still learning about exactly what it means to age with the condition, and what the typical challenges and needs are for people in their middle and older years.

From working with our members over many years we know that although the range of impairments and their severity in spina bifida is broad, and each person's experience is unique, there are commonalities in many of the challenges faced in life and health. Through this survey we aimed to capture and quantify some of the biggest issues we know people in the UK with spina bifida experience as they age.

1.2 Methods

1.2.1 Survey design

From Shine's years of experience supporting adults with spina bifida we have the necessary knowledge of the many health issues people with spina bifida face over their lifetime. We also know that most adults do not get specialist health care from clinicians with expertise in the condition. This knowledge helped shape the topics we wanted the questionnaire to address.

The survey comprised 77 questions and was divided into the following sections: About you; About your spina bifida; You and driving; You and work; Your living arrangements; Your mobility and movement; Your medical history: skin and tissues; Your medical history: Chiari II; Your medical history: tethered cord; Your medical history: kidney, bladder, and bowels; Your mental health; Sleep apnoea; You and pain; Sex-specific questions (Females/Males only); You and healthcare; Hospital admissions. A final page allowed participants to give their contact information to be added to a database of people interested in future research opportunities.

Most questions were multiple choice, but spaces were also provided for free-text input where we wanted to gain qualitative data through respondents' personal insight and experience.

1.2.2 Collecting responses and preparing for analysis

We calculated that 997 was the minimum number of responses needed to be able to extrapolate our results to the wider population of people with spina bifida in England, Wales and Northern Ireland (see Appendix 1). We therefore set a target of 1000 responses, and this informed the name of the survey, the campaign, and the recruitment approach. Participant recruitment was achieved via a social-media campaign, and by postal and email invitations.

All Shine members with spina bifida over 25 were encouraged to complete the study. The first version of the survey went live in July 2019 and ran for 4 months. Based on member feedback, we adapted the online survey to include skipping logic, re-releasing in April 2020 and running for 4 months. Between the online survey releases, we sent out hardcopies of the survey by post to all relevant Shine members. Responses from returned postal surveys were manually inputted via the survey weblink.

The total number of survey responses was 1281, and after removing duplicate and incomplete responses, 1082 surveys remained for analysis.

1.3 Results and discussion

The raw data are presented along with significant results identified by stratification and comparative analysis using a standard 95% confidence interval.

1.3.1 Demographics

The age distribution of respondents matched our predictions based on birth prevalence and survival data: a roughly Gaussian distribution around a peak at ages 46-55 (Figure 1). As we would expect, based on the increased prevalence of spina bifida in women (Källén et al., 1994), 64% of respondents were female, and 35% were male. Most responses (82%) were from members in England, 7% were from Wales, and 10% were from Northern Ireland.

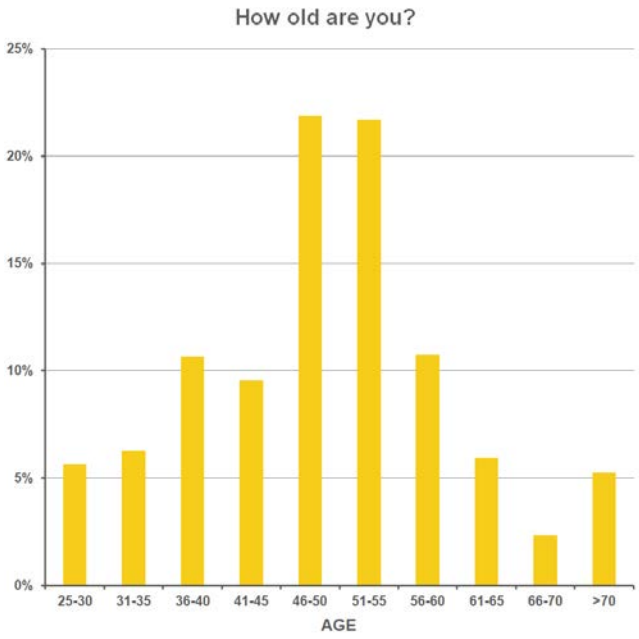


Figure 1: Distribution of the ages of respondents

These demographics, and the fact that we exceeded the minimum responses needed (>1000), gave us confidence that we could draw reliable conclusions about over 25s with spina bifida in England, Wales, and Northern Ireland.

1.3.2 Lesion level

Spina bifida lesions are predominantly found in the lumbar and lumbosacral region of the spine (Riddick-Grisham and Deming, 2011), accordingly the majority of respondents (31%) said their lesion was between their waist and bottom, or close to their bottom (23%, Figure 2). Only 15% knew the exact position of their lesion, and of these 75% were L1 or lower.

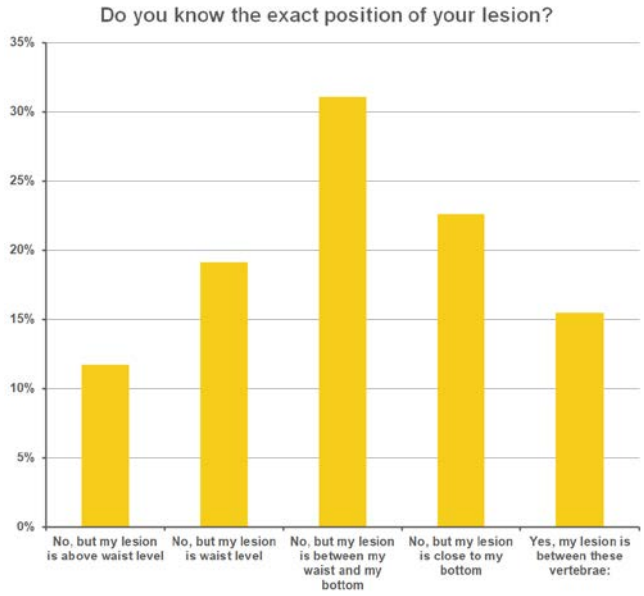


Figure 2: Lesion levels in survey respondents

When the survey results were stratified according to lesion level, some significant differences were apparent but in many respects the health issues were universal. The significant differences seen were:

People with lesions close to their bottom were:

Less likely to have reported worsening spinal curvature over time (42% compared with 61%, 58%, and 59% for lesions above the waist, waist level, and between the waist and bottom respectively)

More likely to have reduced kidney function (48% versus 31%, 38%, and 36% for lesions above the waist, waist level, and between the waist and bottom respectively)

People with lesions above the waist were:

Less likely to have held a full driving licence (56% versus 71%, 66%, and 72% for lesions at waist level, between the waist and bottom, and close to the bottom respectively)

Less likely to have had a pressure sore (45% compared with 64%, 57%, and 57% for lesions at waist level, between the waist and bottom, and close to the bottom respectively)

More likely to have been diagnosed with sleep apnoea (18% compared with 9%, 10%, and 6% for lesions at waist level, between the waist and bottom, and close to the bottom respectively)

(Males only) More likely to have been diagnosed with prostate enlargement (15% compared with 6%, 5%, and 8% for lesions at waist level, between the waist and bottom, and close to the bottom respectively

Section summary

People with spina bifida lesions of all levels experience many of the same health issues. Regardless of lesion level patients with spina bifida must have access to appropriate healthcare, and be helped to understand how to manage and age well with their condition.

1.3.3 Hospital experiences and emergency admissions

53% of respondents had experienced at least one emergency admission to hospital over the last 5 years; 14% reported multiple admissions. The

majority of admissions were for under one week but some were considerably longer, for many months. The average hospital stay for those admitted was 20 days (Figure 3).

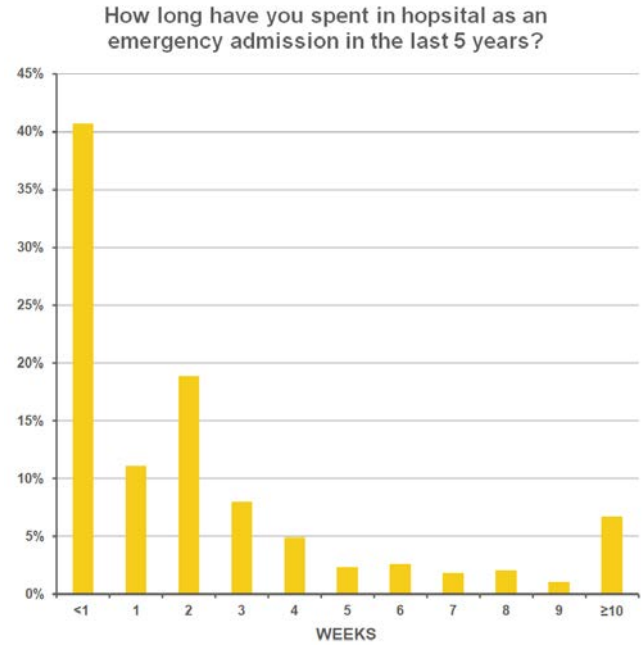


Figure 3: Time respondents had spent in hospital as an emergency admission in the last 5 years

The ten most common reasons for emergency hospital admissions over the last 5 years were: skin and tissues; urosepsis; lung problems; GI problems; shunt problems; UTI; sepsis; kidney problems; and cardiovascular problems. Together these accounted for 64% of all emergency admissions reported (Figure 4 opposite on page 11).

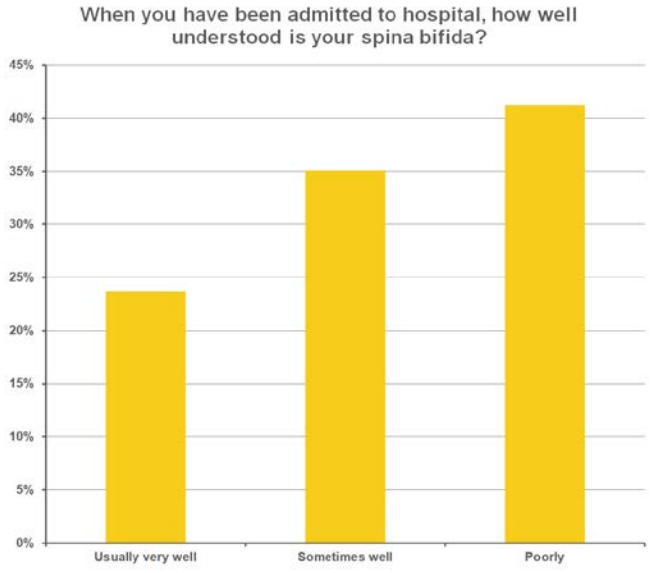


Figure 5: Respondents' experiences of how well their spina bifida has been understood during hospital admissions

41% felt their spina bifida was understood poorly when they have been admitted to hospital, 35% said it was sometimes understood well, and only 24% thought their condition was always understood well (Figure 5).

Our members reported various difficulties occurring during their admissions: while in hospital 43% found it difficult or impossible to get specialist equipment such as pressure-relieving mattresses (Figure 6A), 23% had acquired a pressure sore while in hospital (Figure 6B), and 54% found it difficult or impossible to stick to their bowel management schedule (Figure 6C).

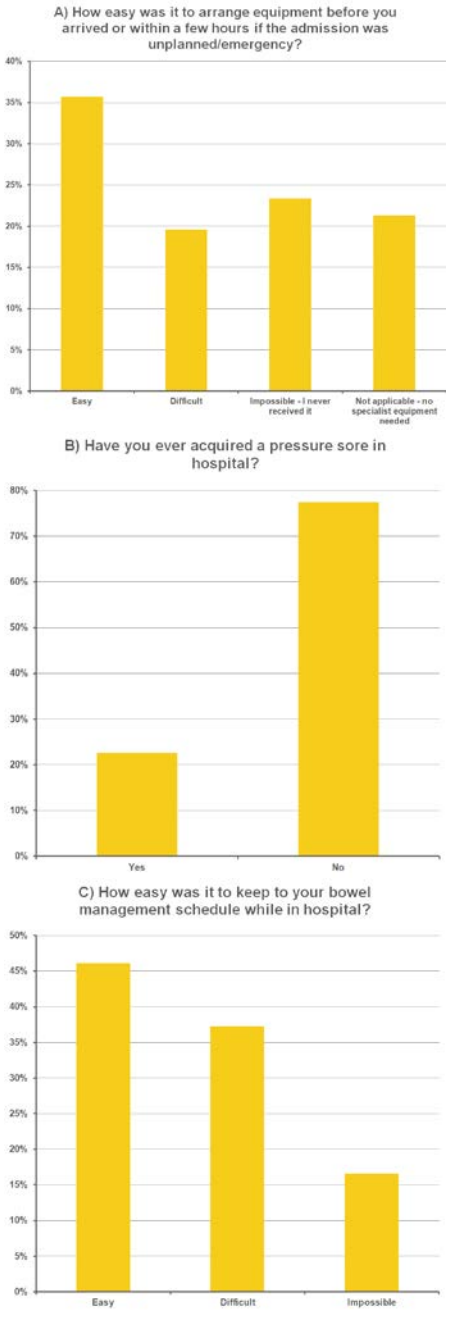


Figure 6: Respondents' experiences around: A) obtaining specialist equipment upon admission to hospital, B) acquiring pressure sores while in hospital, C) on the ease of keeping to their usual bowel management schedule during the stay

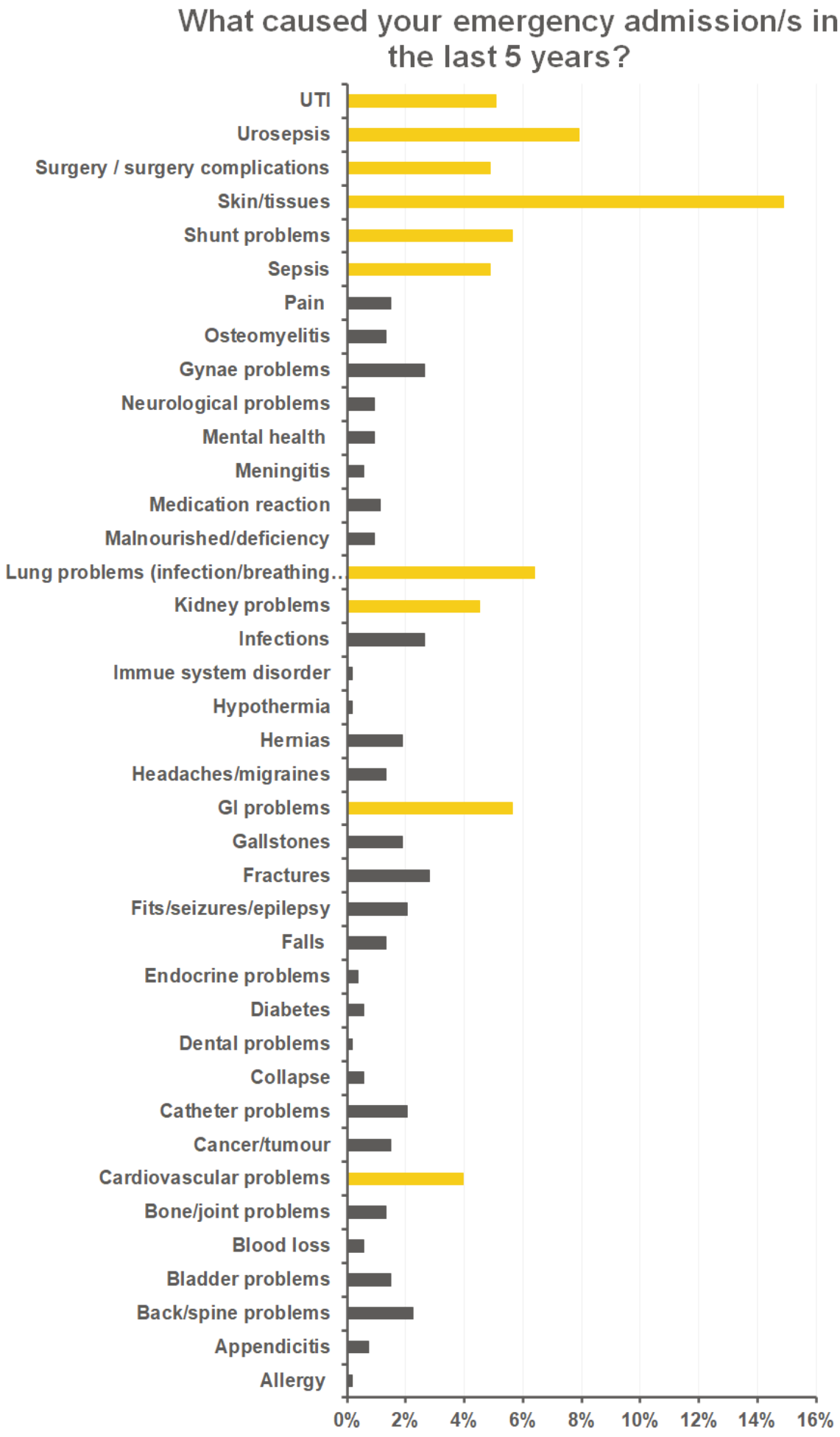


Figure 4: Reasons for emergency admissions in the last 5 years

Throughout the survey respondents shared comments and personal stories about the healthcare they have received and sadly a great many reflected very difficult experiences and negative views (Figure 7). The primary issues related to:

- Lack of understanding, interest, and specialist knowledge of healthcare professionals, particularly GPs
- Receiving reactive rather than proactive care to the detriment of health
- Delays and difficulties accessing treatment and specialist care, physiotherapy especially
- Having to pay for private consultations and care, extra expense
- Low standard of care
- Lack of support and advice, particularly for health needs associated with ageing

“Under no specialist - seems once ur of a certain age u dont warrant”

“nobody seems interested, especially GPs”

“lack of support from NHS funding and no physio”

“I wish I could afford a wheelchair because using a cane now means I barely leave the house”

“I have had trouble finding a physio who isn’t terrified of spina bifida”

“Poor GP services identifying other medical condition too late”

“Have been to see GP, just dismisses anything”

“I feel that I have been left to deal with this myself and any aids I have, I have had to buy them myself, even my GP doesn’t know enough about spina bifida and I have often been sent to see the wrong consultant when I have need to see one”

“Since going into a chair full time my legs have become more bent up & I have developed a left sided partial dislocation of my left hip. I rely on my mum for physio as I can’t get any help on the NHS”

“I feel physio would have kept me mobile”

“I have had private consultations … because of the complexity of my situation. Also because of severe pain I needed to see a specialist quickly”

“Never ever been helped apart from medications that do not help. A x-ray showed scoliosis and nothing since. No help or advice during my whole life”

“infected pressure sore -10 weeks on local hospital -poor care -no daily bowel care -caught chest infection ward too small. Dietrie problems 2 weeks on university hospital now settled for daily home care”

“Surgery on my right shoulder. Rotator cuff. 3/4 tendons were torn. Left for 18 months due to the GP refusing to refer me on the instruction of a pain management consultant. No longer able to transfer as a result”

“Mobility is something important and in my opinion I went with no input as soon as I became an adult. As a child mobility was monitored at children’s clinic. Then zero input once turning 18”

“I’ve not had a check up as regularly as in the past. Have also suffered with pressure sores that unfortunately had to spend a long time at hospital”

“Only specialists I have seen were when I was a child”

“Mostly doctor Do not see specialist regularly. Could be years”

“I have to push for help which was given to me as a matter of course when I was at a special schoolboy after leaving, I was left to navigate my way via the GP to specialists but specialists don’t want to help anymore as I don’t walk, I feel extremely let down”

“am seeing neurosurgeon and ortho consult but only because of need to find out why things are getting worse - has not been a regular thing and only instituted by me because of problems I now know are caused by tethered cord”

“I was only seen by a urologist once the cancer was found which I think is disgusting. All people with my level of paralysis should be under a urologist automatically”

Figure 7: Respondents’ thoughts about, and experiences of, UK healthcare

Section summary

Approximately half of adults living with spina bifida will need at least one emergency admission in 5 years. These admissions can be lengthy (on average 20 days) so they have high financial as well as human costs. Patients generally felt their condition was not well understood or accommodated by healthcare services and many had struggled with access to regular, specialist care. Spina bifida is a complex condition with a host of associated healthcare issues. To help patients better meet the challenges of ageing with spina bifida and improve their healthcare experiences we recommend: regular, proactive checks; greater involvement and communication between specialists; improved knowledge and understanding of spina bifida by all professionals involved in their care.

1.3.4 Independent living

1.3.4.1 Driving

Driving is an important facet of independence in adulthood. In England, 75% of adults in the general population have a full driving licence and the percentage increases with age from 62% aged 21-29 up to 85% in those aged 60+ (ONS, 2020). 67% of our survey respondents had at some time held a driving licence, and the proportion generally increased with age from 42% at 25–30 to 80%+ in those aged 66 and older. This suggests that while the percentage of licence-holders with spina bifida is slightly smaller in the younger adults, over the years the number of licence-holders catches up to the general population. Of those who had held a full a licence, 81% were still driving, and 68% of those drive anywhere they choose.

The most common age ranges to have stopped driving were: 36–40 (18%) and 21–25 (15%) and the median age to stop was 38. People who had stopped driving did so for a range of reasons including: concentration/cognitive difficulties, anxiety/depression, physical/movement changes, financial reasons, loss of sensation/weakness in the legs, medication, and seizures.

1.3.4.2 Living situation

In 2019 the Office of National Statistics reported that in the UK 8.2 million people (16% of the adult population) were living alone, and 19.2 million were living with family (37% of the adult population). 26% of 20–34-year-olds in the UK were living with their parents (ONS, 2019b). A variety of living situations were also reported in our survey; the proportions were similar though a greater percentage were living alone. We found that 32% of respondents lived alone, 22% with their parents, 38% with a partner, and 9% with children.

The proportion of those living alone increased with age from 6% aged 25–30 to a peak of 48% in the 66–70 age range. The period of time people had lived alone varied greatly from 1 to 50 years, the most common length of time living alone was 16–20 years (21%), and the median duration alone was 17 years. By stratifying the results according to living situation we found that living alone did not significantly impact on health outcomes.

A much larger proportion (68%) of those aged 25–30 lived with their parents compared with other ages, the next highest being those aged 31–35 (38%), and the proportion declined further with advancing age. More men than women lived with their parents (27% versus 20%), and more women than men lived with their children (11% versus 5%). The three main reasons given for living with parents were: being happy there, having high care needs, and financial reasons.

1.3.4.3 Work

In 2019 the UK employment rate across all working-age adults was estimated at 76% (ONS, 2019a). Employment was highest among adults aged 35–49 (85%) and 25–34 (84%); employment in 50–64-year-olds was 73%. Across all survey respondents, 61% reported not currently working, 12% were in full-time paid employment, 15% were in part-time paid roles, a further 5% reported working but did not specify hours or pay status. Because our survey included people of and above retirement age, we also examined employment rates in respondents 65 and under i.e., working-age. The proportion of 25–65-year-olds not currently working was 59%, 13% were in full-time paid employment, 16% were in part-time paid roles, and 6% were working but did not define hours/pay further. These rates are similar to the rates across respondents of all ages, though

there is a slight increase in employment.

The proportion of people not working generally increased with age from 52% at 25–30 to 70%+ when over 56 (Figure 8). When stratified by biological sex, more men work in full-time paid roles than women (16% compared with 10%), and more women than men work in part-time paid employment (17% compared with 11%).

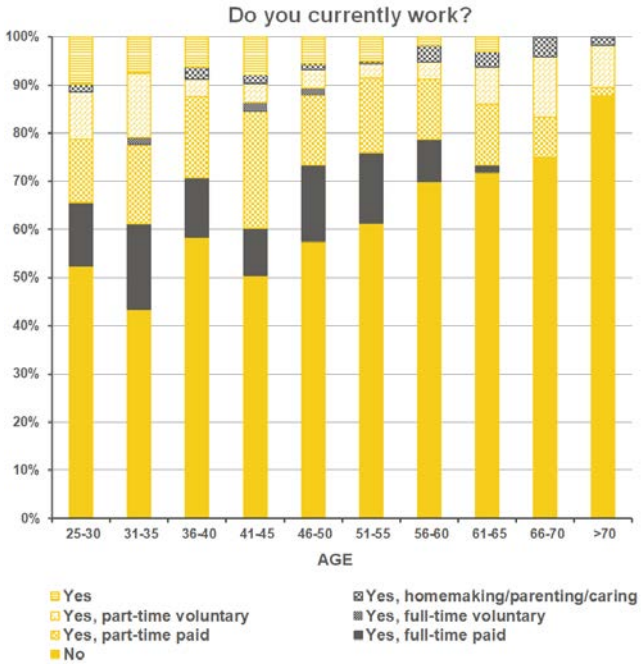


Figure 8: Working situation of respondents in different age brackets

56% of respondents said their work situation has changed over time and the proportion increased with age, reaching a maximum of 76% at ages 66–70. The most common changes in work situation were: from working to not working; reduction in working hours; changes from paid to unpaid work; and changes staying within paid work (Figure 9a).

The most common age ranges for changes to occur was 36–40 (22%), 46–50 (16%), and 26–30 (14%). The median age for work changes to occur was 40.

The most common reasons for changes in working situation were: pain, bladder and bowel issues, anxiety/depression, health deterioration, starting a family, retirement (early/medical and at retirement age), redundancy, loss of mobility, issues with employers, and being unable to cope (Figure 10).

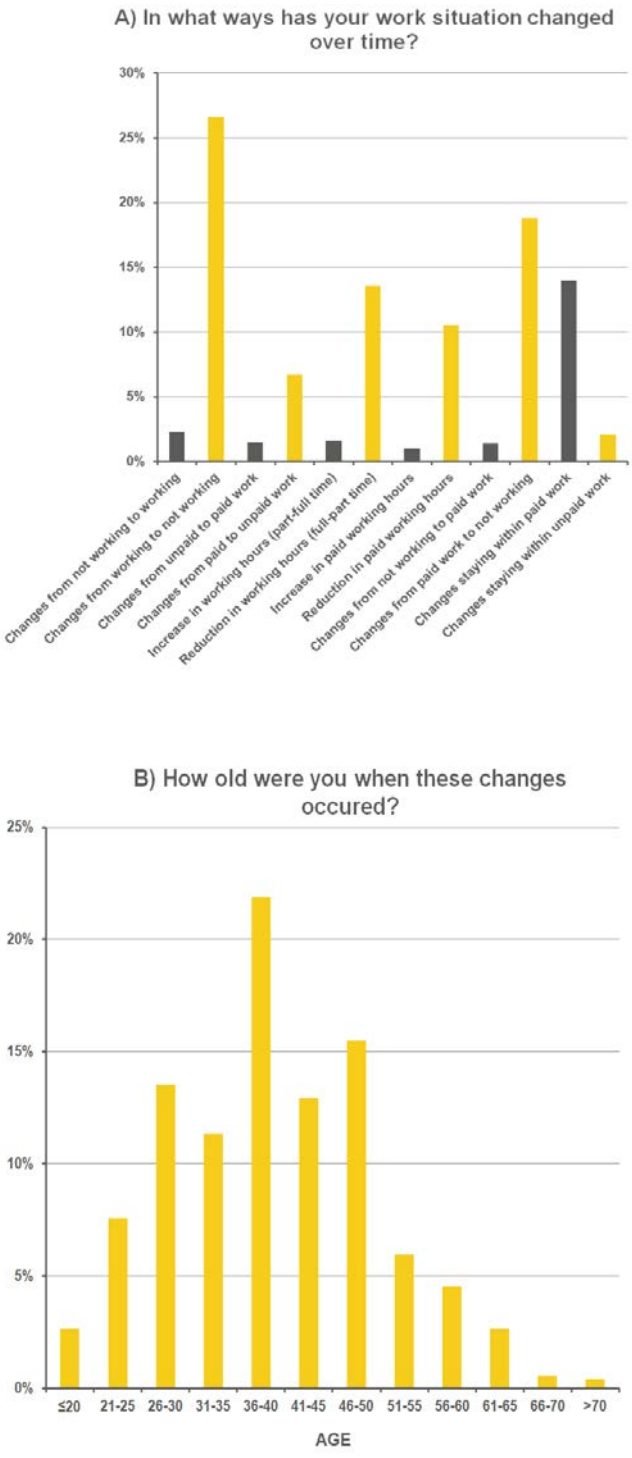


Figure 9: Answer classifications and responses to Question 13 – How has your work situation changed over time? and Question 14 – How old were you when these changes occurred?

> *"I was about 38 when my health declined further making it impossible to continue (My health day to day being so unpredictable). This meant I was unable to commit to anything."*

> *"I was 'dismissed due to health problems' then given ill-health retirement. Mostly due to back and breathing problems often causing hospital admissions."*

"I was effectively laid off when employment services' financial support to my employer ceased"

"redundancy. I've had two work capability assessments + two tribunals (one failed/one won) both done by people who were incapable of understanding my situation"

"I went from full time paid to part time after my amputation in 2014 then back to full pay in 2019 due to PIP being lowered and not getting any financial help elsewhere"

"Job description changed did not have the strength to do it. Had to take redundancy."

"Went freelance for greater flexibility, tiredness, lower pressure work, less international travel"

Figure 10: Why respondents stopped working – in their own words

Section summary

People with spina bifida can lead independent lives: living, driving, and working as they choose. Independence is threatened by the worsening and/or accumulation of multiple health issues with age. Proactive condition management and early intervention with health issues can help ensure patients live more independently and have a better quality of life for longer.

1.3.5 Mobility and movement

The ability to move and travel without assistance from other people, but where needed with the use of mobility aids, is crucial for being able to participate fully in life activities and for living as independently as possible.

The need for different mobility aids varies between individuals with spina bifida based on a number of factors relating to body structure and function e.g., lesion level, weight, skin integrity, presence of hydrocephalus, joint pain/stability, general health, visual impairment, balance.

Other external factors also influence mobility including surroundings, and access to suitable equipment and specialists such as physiotherapists and occupational therapists (Vladusic and Phillips, 2008).

17% of respondents reported walking unaided everywhere, and 16% walk with sticks and crutches all the time. 25% of respondents use a manual wheelchair all the time, 6% use one but stand to transfer, 7% use one for long distance only, and 8% for outdoors only. 5% reported using a mobility scooter outdoors and 15% use a power chair. The most common age range to start using a power chair was 41–45, 70% began using their power chair at 45 years old or under, and the median age to start was 37 (Figure 11).

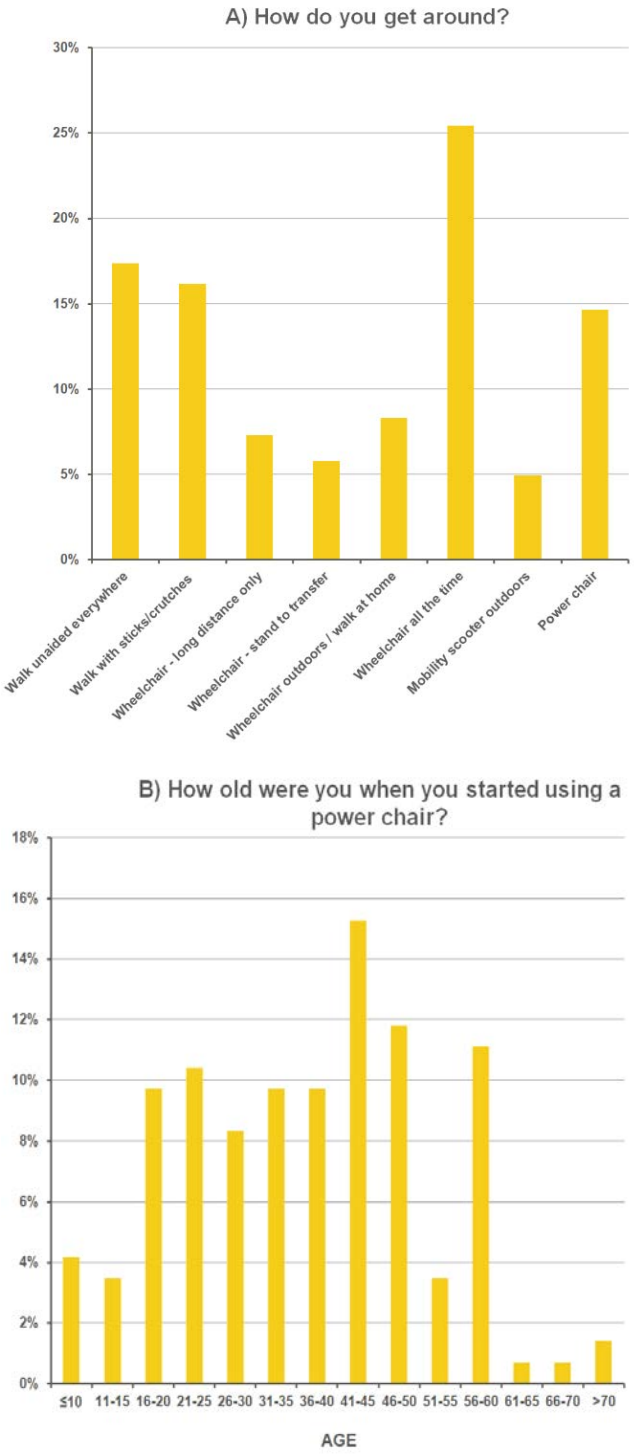


Figure 11: A) Mobility of respondents. B) Age power chair users began using this mobility aid

65% of wheelchair users had used a wheelchair for over 20 years, and the median length of wheelchair use was 30 years. 34% of those who use sticks/crutches had done so for more than 20 years and the median length of use was 15 years. 75% of respondents said their mobility has changed over time, and when stratifying by age, the proportion of those reporting a change in mobility increased from 48% in 25–30-year-olds to 90% in those aged 71+. A wide variety of changes in mobility over time were reported (Figure 12) but these changes can be broadly divided into those representing a decline in mobility (78%), improved mobility (13%), and changes best described as differences (9%).

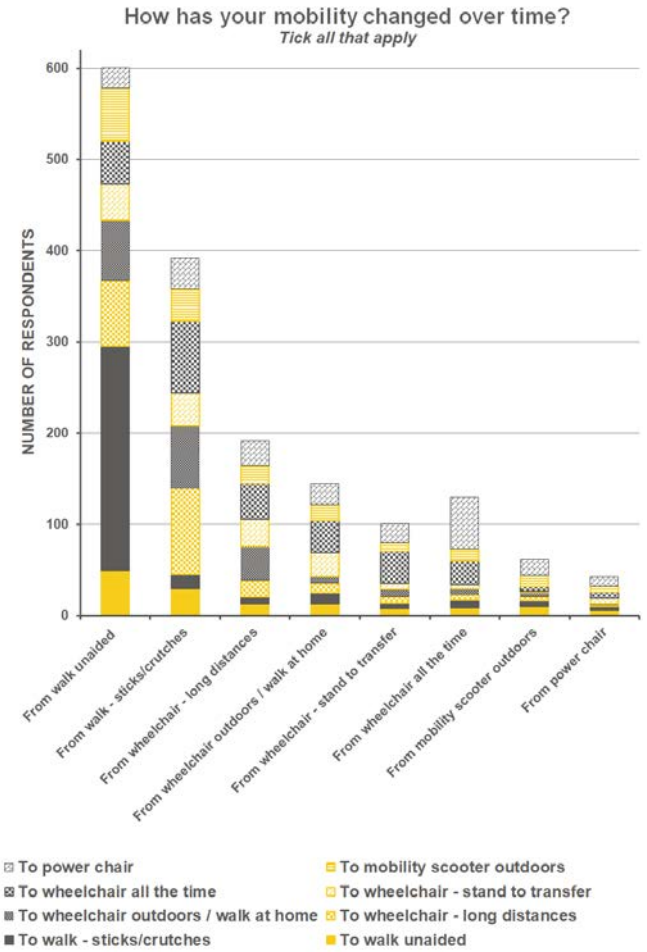


Figure 12: Changes in respondents' mobility over time

Changes in mobility were reported for a broad range of ages between 11 and 50 years old, the most common was 36–40 (14%), and the median age for changes to occur was 35 (Figure 13). The reasons given for mobility changing were varied but frequently cited: pain, balance problems, age, deterioration, surgeries, arthritis, sensation loss, weakness, falls, weight gain, and pressure sores.

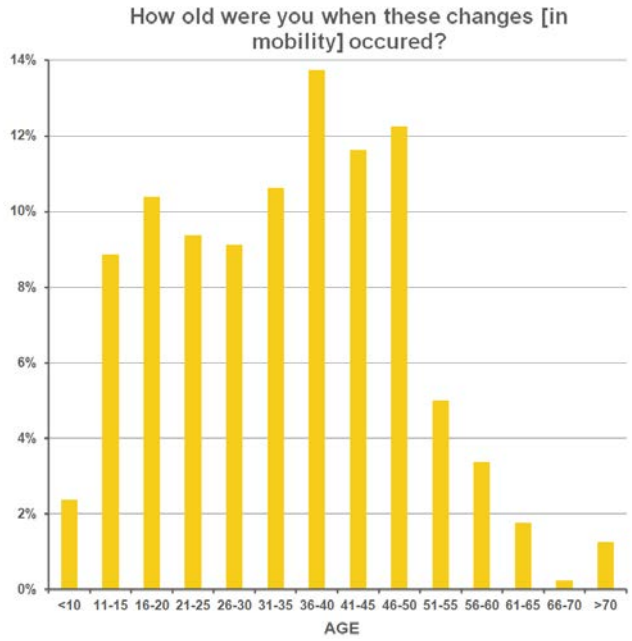


Figure 13: Ages when respondents' mobility changed

We gave respondents space to add their own comments about their mobility and the range of views and experiences shared reflects the complexity and diversity of spina bifida and those affected by the condition. Examples are given in Figure 14. Common themes and experiences were evident from the free-text comments: mobility deteriorating with age; frustration with the changes and wishes to have been better prepared; the mutual impact of mobility and other health aspects including weight, pain, sensation, and continence; the benefit of exercise; the pros and cons of wheelchair use (independence versus upper body issues and weight gain).

> *"I was stable for most adult life I had no concept that things would change/worsen"*

> *"the most significant reduction in mobility has been since 50. the deteriation has been quite profound and has had an impact upon my life"*

> *"I feel too young. To be walking with aids, my spina bifida, got a hold of me, bringing nerve damage, falls, incontinence of bladder and bowel. I can't accept seeing myself with walking aids"*

> *"Although I still walk unaided I do have more pain when walking and I'm not as stable as I used to be"*

> *"Upper body too heavy for legs as I aged; elite athlete - used chair for speed /convenience/laziness"*

“It was easier to get round with a wheelchair rather than calipers + sticks and I felt less disabled as people didn’t stare as much”

“Mobility affected by Pain, numbness, spasms, cramps, bladder and balance”

“I wish I had continued to walk and not relied on wheelchairs. Once using chairs I gained lots of weight My weight gain has dramatically effected my mobility”

“I enjoy using a wheelchair, it’s fun! I can shop, do sports, carry items on my lap, hold hands with boyfriend/grandchildren when out, cook, play with grandchildren, and I’m still very sexually active at 60, dating a police officer aged 45! Generally be independent!”

“I have good days and bad days. Bad days are very stiff and a hey in my back and legs, moving and walking is an effort”

“I hate not being able to move so well. I do go to a physio gym class which helps a lot. I find i get other things to slow me down now, bladder problems, dizziness etc”

“I have been to physiotherapy in the last year and lost a lot of weight and have gotten a lot of mobility back. I am now walking with crutches more often and sometimes outside”

Figure 14 (above and opposite): Respondent comments on mobility

56% of respondents reported a reduction in leg movement since turning 18, when stratified by age the trend showed an increase from 32% at 25–30 to 70–83% in those over 61 (Figure 15). When asked the age at which the movement in their legs reduced, most answers (57%) fell between the range 21–40; the median age was 35.

We asked our members if they’d had joint replacements: overall most had not (91%). Stratifying by age reveals that the proportion who’d had a joint replacement increased with age from 6% on average for the 50 and under to 22% on average for the 51+. Females were more likely to have had a joint replacement than males (13% compared with 7%).

70% of respondents said their balance has worsened over time and this trend was reflected in the age-stratified results where the percentage reporting worsening balance increased from 32% aged 25–30 to between 80% and 94% in people 61 and over (Figure 16). Women were more likely than men to report increasing difficulties with balance (74% compared with 42%).

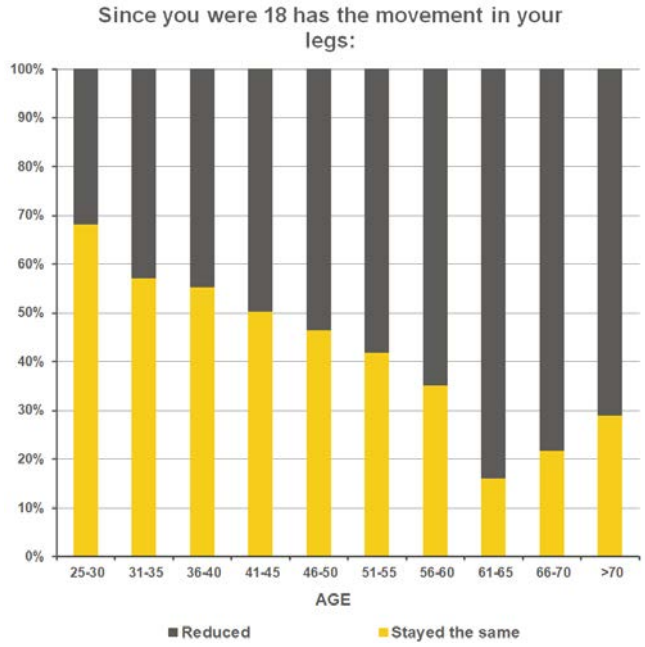


Figure 15: Changes in respondents’ leg movement with increasing age

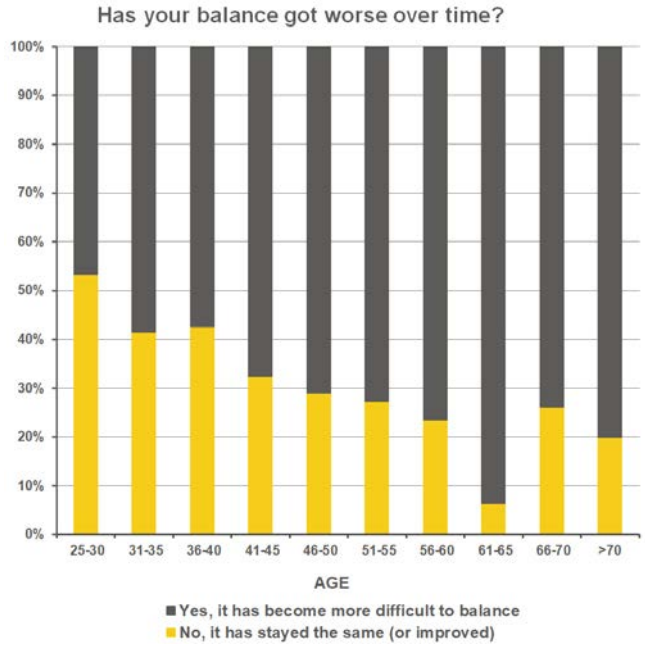


Figure 16: Changes in respondents’ balance with increasing age

Only 31% of respondents reported no upper body problems; 45% had two or more issues. Shoulder pain was the most common upper body issue with

53% of respondents experiencing this. 48% had neck pain, and 33% had difficulty using their hands (Figure 17).

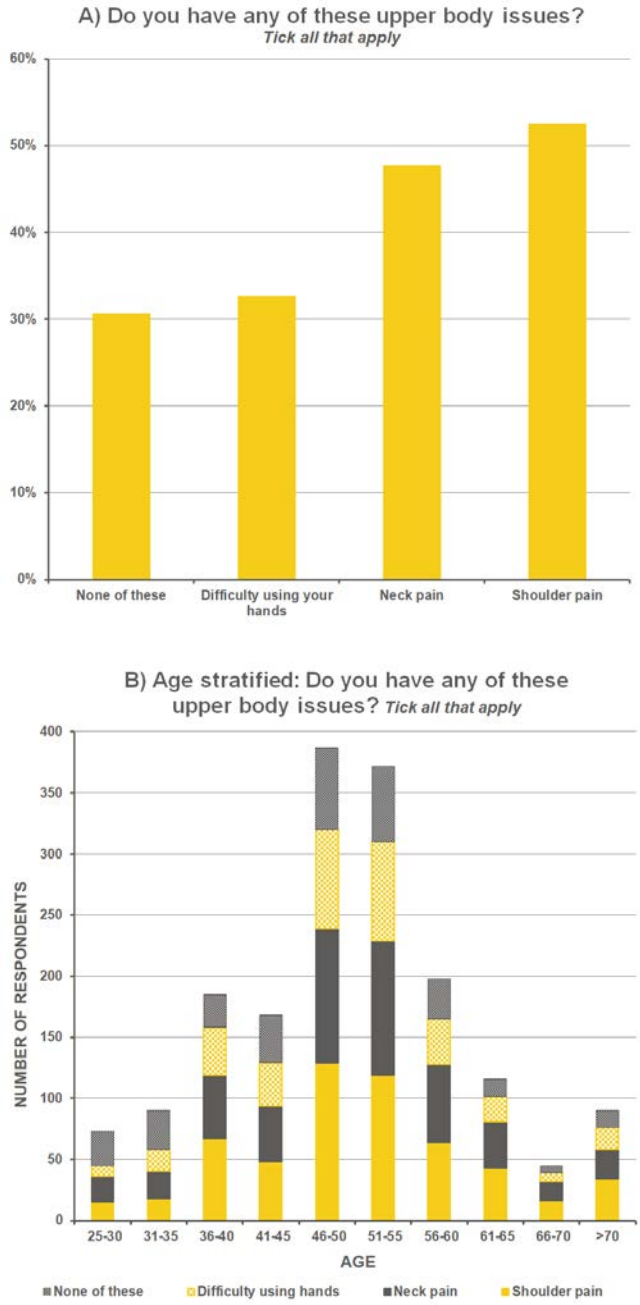


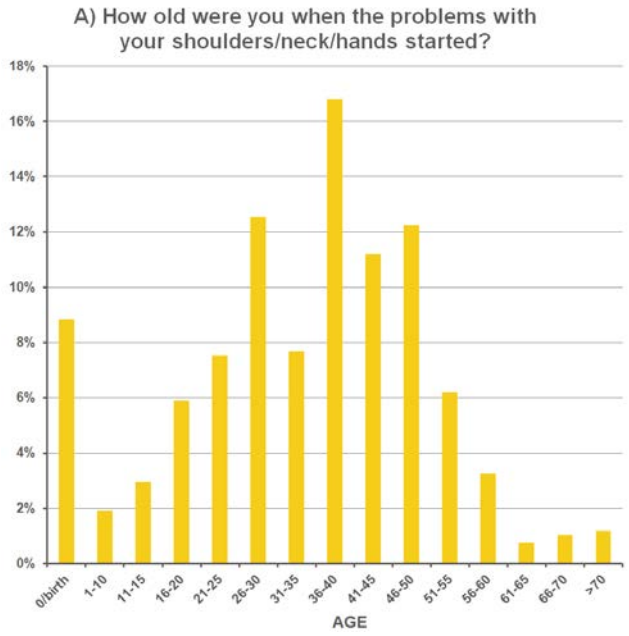
Figure 17: Upper body issues experienced across A) all respondents B) stratified by age

To determine if there were specific mobility aids or approaches that were associated with better outcomes for the upper body, we stratified the upper body pain/difficulties data by the different ways that people got around.

People who walked unaided everywhere were less likely to have upper body issues: 44% reported having none compared with 23–31% for the people who used different mobility aids.

There were no significant differences between the other groups including manual wheelchair use at different levels, walking with sticks/crutches, power chairs and mobility scooters.

The most commonly reported age range for upper body issues to start was 36–40 (17%) and the median age of onset was 38 (Figure 18). Upper body problems hindered respondents in carrying out daily chores and personal care, and affected their independence, ability to transfer, and tasks/hobbies requiring manual dexterity. Some said the pain didn't prevent them from doing anything, but made some tasks more difficult.



> “Makes using crutches harder and more painful. Using wheelchair also more difficult. I don’t go out as much”

> “I also suffer from arthritis bad in my arms and legs and hands now so cannot support myself to stand for long periods of time or to walk anymore”

> “Writing is difficult and painful for me.I get problems with my shoulders which make things like shopping, changing the bed sheets and hoovering exhausting”

> “When my shoulder is at it’s worse I am bed ridden or rooted to the floor in agony. This has happened in some embarrassing places. At it’s worse the pain sends me straight to the floor regardless of what I am doing. This can be difficult when in public or around the house”

> “Every day tasks. I can’t lift items due to the pain it causes. Moving my arm, neck and shoulder causes extreme pain”

> “My neck and shoulders are painful when walking, or stretching. My right hand has very little strength, which makes manual tasks difficult. I find writing and using cutlery difficult”

> “Makes me feel unstable, struggle doing basic tasks half the time such as dressing, bathing etc”

> “Don’t stop me doing anything just makes it more difficult and everything takes longer”

> “Just make things painfull but I do what I want”

Figure 18 (above and opposite): A) Ages when respondents started experiencing problems with their shoulders/neck/hands B) Free-text analysis of what the upper B) Free text analysis of what the upper body issues prevent respondents from doing. C) Respondents describing the impact of upper body issues in their own words.

Section summary

Deterioration in mobility often begins in early adulthood, between 25 and 40 years of age. This deterioration may be slowed or prevented with regular monitoring and early intervention. The use of any mobility aids is associated with pain and problems using the upper body and hands which impacts on independence and quality of life.

Section summary continued

Patients should have access to occupational therapy assessments and physiotherapy at every mobility transition point (e.g., from walking unaided to using sticks, or from walking with crutches to using a manual wheelchair). Earlier introduction of power chairs may prevent pain and loss of independence.

1.3.6 Bones and joints

Overall, 68% of respondents reported being diagnosed with a bone/joint condition (Figure 19A). When stratified by age a significant trend of bone/joint problems accruing with age was revealed: 52% of those aged 25–30 reported at least one bone/joint issue compared with 75%+ in the people aged 51 and over, and 86% of the 71+ respondents.

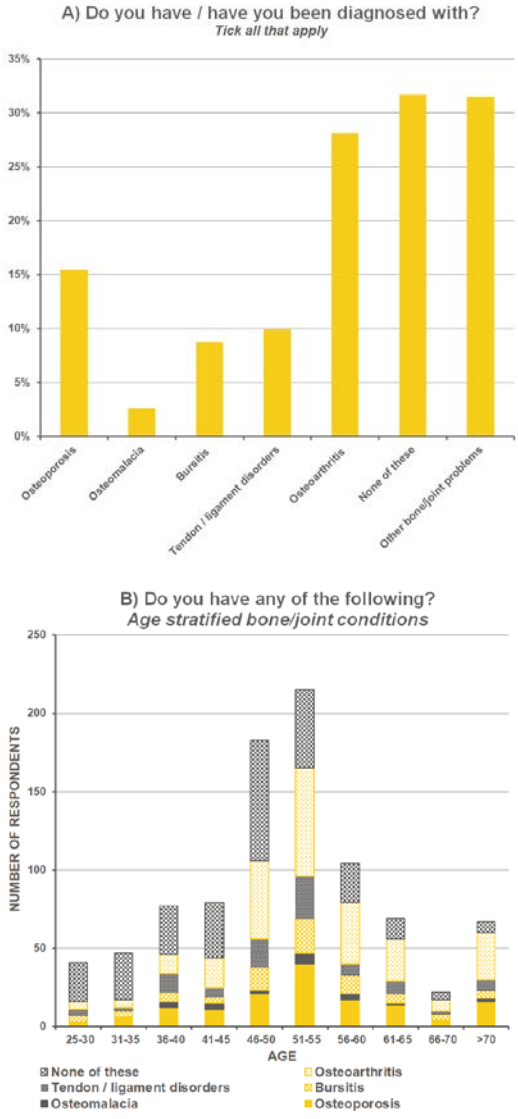


Figure 19: Responses to Question 34 – Do you have/ have you been diagnosed with the following bone/joint conditions? Results for all respondents, and stratified by age, and biological sex

Osteoarthritis is a condition characterised by damaged and lost cartilage causing painful inflammation and stiffness in the joints. The condition typically occurs from accumulated wear and tear over the course of a lifetime, but its progression is accelerated by overuse of the joints or trauma. 28% of respondents reported having been diagnosed with osteoarthritis (Figure 19A), significantly higher than the general UK population of 10% of adults over 20 (Swain et al., 2020). As would be expected, prevalence increased with age (Figure 19B), starting at 9–10% in the 25–35s and rising steadily, eventually reaching 59% in the 71 and over group.

Bursitis is a condition involving painful inflammation of the bursae: the small, fluid-filled sacs that protect the bones, muscles and tendons at the joints. The overall bursitis prevalence revealed by our survey was 9%, but stratification by biological sex demonstrated that women were significantly more likely to report having the condition than men: 11% versus 4% (Figure 19A).

Osteoporosis is a condition characterised by a loss of bone density and strength, known to be strongly associated with age. Bone loss is particularly marked in post-menopausal females because of the role of oestrogen in bone mineralisation. Reduced weight bearing (wheelchair use) and factors such as poor kidney function and medication for epilepsy increase the risk for osteoporosis, and a Swedish study has previously suggested the rate in people with spina bifida was 33%, higher than the general population (Valtonen et al., 2006).

UK osteoporosis prevalence for females at 50 years old is estimated to be 2%, while in those over 50 it is estimated at 21.8%. For males over 50 the prevalence is 6.8% (Kanis et al., 2018; NICE, 2012). 15% of survey respondents had been diagnosed with osteoporosis, and the proportion increased with age from 6–13% in people aged 25–50 to 17–32% in the respondents aged 51 and over. The prevalence in males 50 and under was 7%, rising to 10% aged 51+; the prevalence in females was significantly higher: 10% in those 50 and under and 22% in the women who were 51 and over (Figure 20).

The data suggest that the risk of osteoporosis is significantly raised for people with spina bifida, particularly in groups not routinely checked for bone density e.g., under 50s and males.

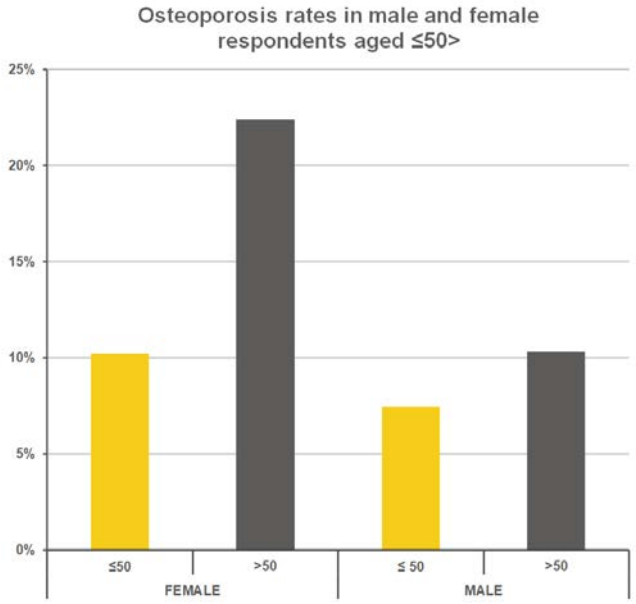


Figure 20: Age- and sex-based prevalence of osteoporosis in respondents

The average annual prevalence of fractures for UK adults is dependent on age, as with osteoporosis this is due to age-related bone loss. The prevalence in those aged 18–49 has been reported as 0.73%, and as 1.19% for people aged 50 and over (Curtis et al., 2016). 18% of respondents aged 50 and under, and 22% of those over 50, had experienced at least one fracture in the past 5 years (Figure 21). This suggests an annual fracture prevalence of 3.6% in adults with spina bifida aged 25–50 and of 44% in the over 50s, i.e., at all ages the fracture risk is above that of the general population. This is likely a result/reflection of the increased risk of osteoporosis with spina bifida.

All spines have some curvature to aid balance and movement. Atypical curvature of the spine, such as scoliosis, is a known complication of having spina bifida. Disorders of spine curvature disrupt balance, impair mobility, and can increase the risk of pressure sores due to uneven weight distribution. 57% of survey respondents reported having curvature of the spine and it was more common in females (62%) than males (50%).

There were no significant differences in the proportion of people reporting curvature of the spine when stratifying by age, however 57% of people with curvature of the spine felt that it had worsened over time. This suggests that curvature of the spine develops before the age of 25 and worsens with age.

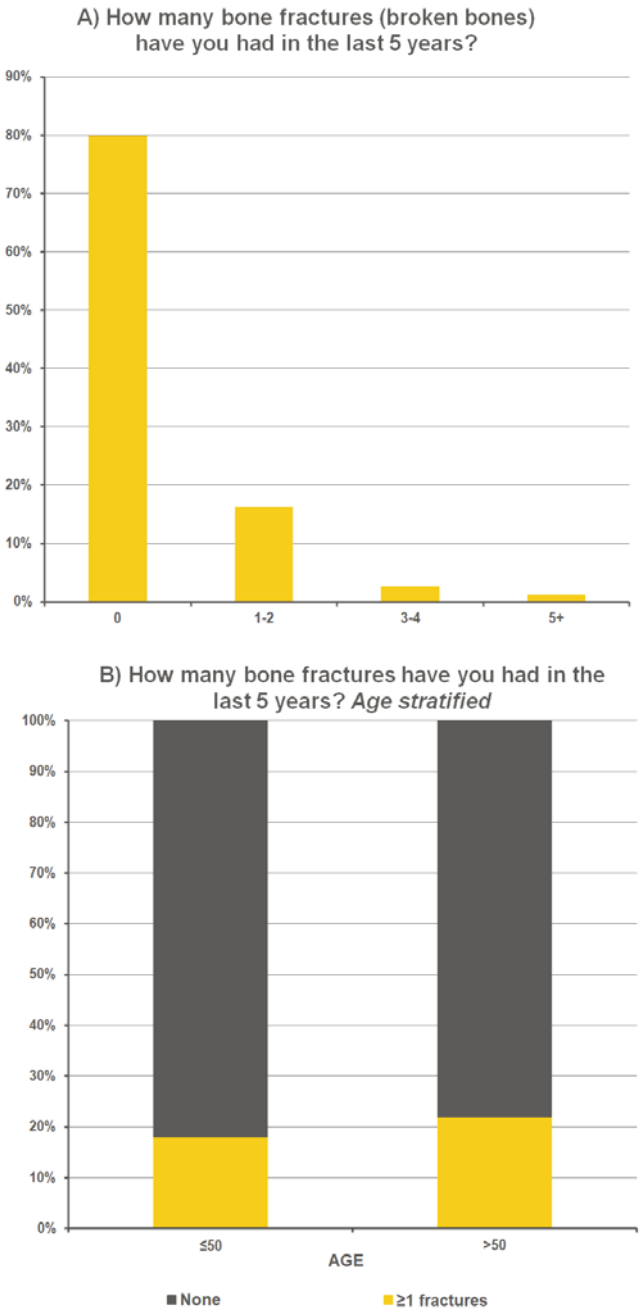


Figure 21: Responses to Question 35 – How many fractures have you had in the last 5 years?

Section summary

Having spina bifida significantly increases the risk of osteoporosis and osteoarthritis. To help prevent and manage osteoarthritis suitable mobility aids must be introduced at the right time, exercise/ physiotherapy encouraged/provided, and better management of pain and weight is needed. Underdiagnosis of osteoporosis is a problem that may be addressed by bone density screening for men and women with spina bifida throughout their adult lives, and beginning in their twenties.

1.3.7 Skin and tissues

Cellulitis is a condition where a bacterial infection spreads through the deeper layers of the skin, most commonly in the lower limbs. Risk factors for developing cellulitis include obesity, lymphoedema, skin ulceration and the biggest predictor is having previously had cellulitis, with annual recurrence rates estimated at 8–20% (Atkin, 2016). Left untreated cellulitis can cause serious complications, as infection can spread into the muscles, bones, and bloodstream. Cellulitis affects approximately 2.5% of people in the UK annually (Simonsen et al., 2006). 35% of survey respondents said they had experienced cellulitis. The most common age range to have had cellulitis was 21–35, and the median age was 35 (Figure 22). This is younger than the typical age of onset in the UK, which is between 40 and 60 years old (Atkin, 2016).

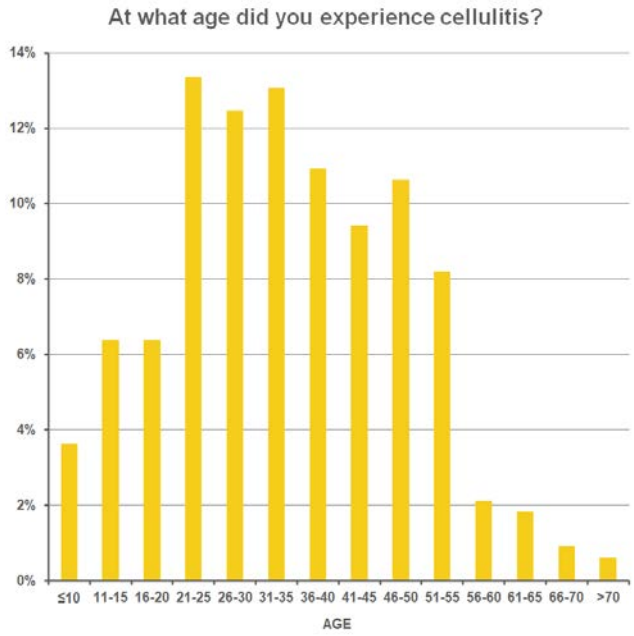


Figure 22: Ages at which respondents experienced cellulitis

Lymphoedema is a chronic swelling of the tissues that occurs due to inadequate fluid drainage by a damaged or dysfunctional lymphatic system. Lymphoedema is often caused by immobility, as the movement of muscles around the lymph vessels helps to squeeze fluid through them. Movement is therefore an important component of drainage function. Lymphoedema affects between 0.13% and 0.40% of people in the UK, and prevalence increases with age to 1.3% of those aged 65–74, and 2.8% of those over 85 (Moffatt and Pinnington, 2012).

We found that 34% of respondents had experienced lymphoedema; the proportion largely increased with age from 20% in 25–30-year-olds to 43% in those aged 56–60. Interestingly, in the respondents aged 61 and over the proportion did not continue to increase and instead declined to 28% having had lymphoedema.

Most respondents reported experiencing lymphoedema at ages 26–30 (13%) and 36–40 (13%). The median age of onset was 35 (Figure 23). Of those who had experienced lymphoedema, 45% had received compression treatment.

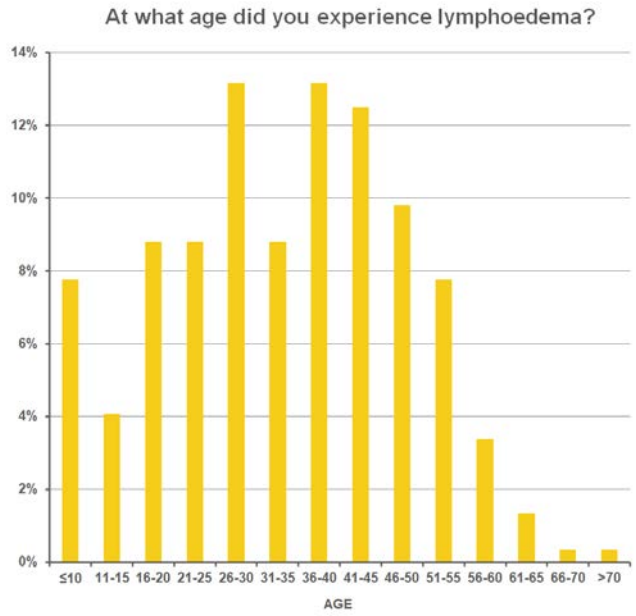


Figure 23: Ages at which respondents experienced lymphoedema

When asked how lymphoedema affects their lives, the main issues highlighted by respondents were: pain/discomfort; difficulties moving legs and in transferring; mobility issues; and problems finding suitable, comfortable footwear.

The mental health impact of lymphoedema was evident, as members reported feeling self-conscious about their legs, and isolated due to the impact of the condition/treatment on their ability to go out and socialise, shop, etc. (Figure 24).

> *"swelling in in legs hard too move them and transfer"*

> *"mobility issues. difficulty buying shoes, wearing socks, toileting as leg is heavy to move"*

> *"Reduces my social life due to being stuck in one place for long periods of time"*

> *"I take diuretics which can limit where I go in the morning as I need to be near a toilet"*

> *"I manage to now keep it under control with diet and minimal exercise"*

> *"Makes me miserable and fed up"*

“Legs are heavy and clothes don’t fit. It’s also embarrassing. Hands swell when I’m walking and they’re down by my side for a while”

“It can make transferring from my wheelchair difficult - heavy real leg legs. I’m often unable to lift my legs. When my Lymphodema us uncontrolled it seems to produce blisters & uncle’s that are hard to heeled (which reaccures)”

Figure 24 (above and opposite): How lymphoedema affects the lives of respondents

Pressure sores (also known as pressure ulcers or bedsores) are areas of the skin and underlying tissue that are damaged by prolonged/recurrent pressure or friction on the skin.

People who use wheelchairs or orthoses and/or who have sensation loss are particularly at risk of developing pressure sores. 55% of survey respondents had had a pressure sore, and they were more common in males (63%) than in females (50%; Figure 25).

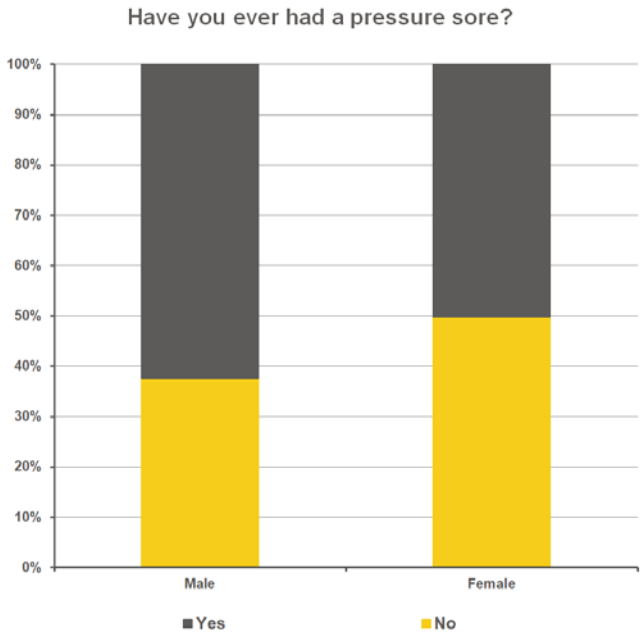


Figure 25: Percentage of respondents who had experienced pressure sores, stratified by sex

When asked the age at which they’d experienced a sore: 16% reported having had multiple sores over the years, including recurrent sores from childhood – some reported having had too many to count.

The most common ages to have sores were 11–15, 16–20, and 26–30; the median age of onset was 25 (Figure 26A). The most common sites for people to have experienced pressure sores were the buttocks (47%) and the feet (44%; Figure 26B).

57% of those who’d had a pressure sore had needed to stay in hospital as a result. The typical duration of a hospital stay was 2–3 weeks (Figure 27) though stays of several months were reported in some cases, and follow-up treatments in the community were described, e.g. “3 weeks [in hospital] then nurse calling at home for 3 months”.

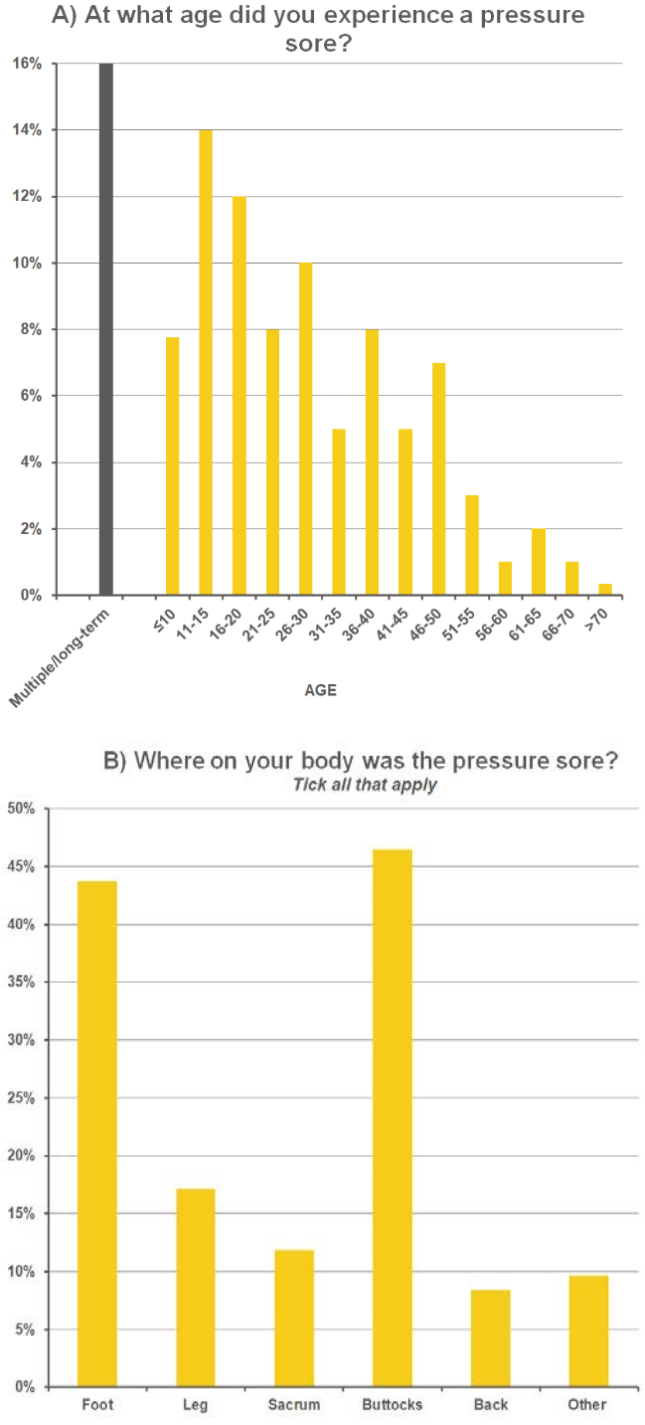


Figure 26: A) Ages when respondents experienced pressure sores. B) Anatomical locations of respondents’ pressure sores

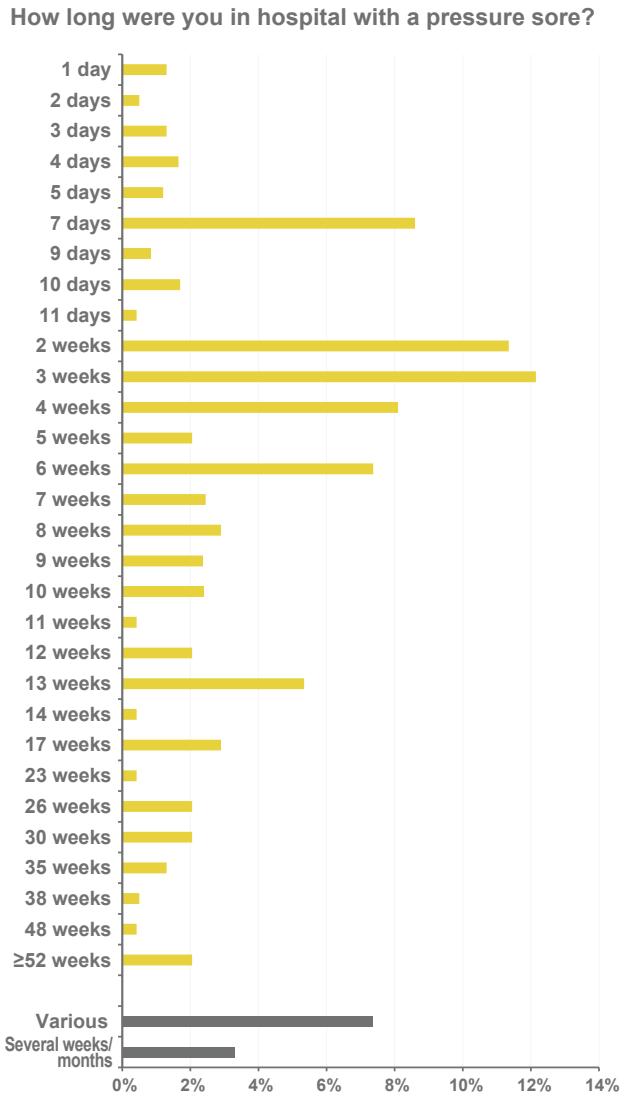


Figure 27: Lengths of hospital stays for respondents who had undergone an emergency admission in the last 5 years

Unprompted, a number of respondents described their experiences with pressure sores (Figure 28). These demonstrated the importance of regularly changing position, diet, and early intervention to prevent escalation to severe infection, sepsis, and the need for surgery – potentially amputation.

> *"I often get them on my heel of my foot when I've been sat in my chair too long"*

> *"1st one when I was 9 years old which lasted about 6 years, had surgery to get rid of it. In 2015 had sepsis from a sore i wasn't aware of until checked by nurse at hospital. Spent 5 weeks in hospital and 5 months in convalescent home"*

> *"I had one at age 29 that took my coxycs bone, due to bad diet, trying vegetarianism and doing it irresponsibly. I was in for 5 weeks and I nearly died"*

> *"Amputation of right leg as a result of pressure sore on the ball of my foot"*

Figure 28: Unprompted comments from respondents describing their experiences with pressure sores

Analysing all the responses for the skin and tissue questions, we were able to determine that only 22% had reported no skin/tissue conditions, 30% reported having one, while 48% reported two or more (Figure 29).

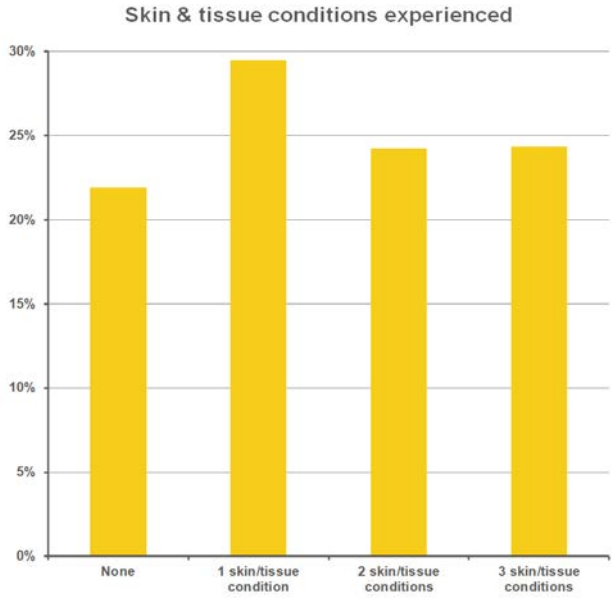


Figure 29: Percentage of respondents reporting skin/tissue problems

Section summary

Loss of sensation, reduced mobility and the use of mobility aids and orthotics increase the risk of pressure sores in people with spina bifida. Cognitive impairment and loss of sensation can reduce the likelihood that help will be sought promptly, meaning pressure sores may go untreated until they are a higher grade and a significant health threat. Early intervention is critical: regular checks to ensure mobility aids and orthotics fit correctly, improved patient education, and prompt access to coordinated, effective treatment.

Lymphoedema is a prevalent issue for people with spina bifida that significantly impacts on independence and self-esteem, and increases the risk of pressure damage. Early intervention with compression treatment, exercise and manual lymphatic drainage is key to minimising the impact of lymphoedema.

1.3.8 Chiari II malformation

Chiari II is a developmental condition of the brain where there is displacement and crowding of cerebellum and brainstem. It is strongly linked with myelomeningocele, the most common form of open spina bifida. Chiari II is a congenital condition (present from birth) but symptoms may not occur until later in life, or may never occur. Typical symptoms of Chiari II in adulthood include: headaches (particularly at the back of the head), neck pain, dizziness/balance problems, numbness/tingling of the limbs, weakness/paralysis of the muscles, hearing loss/tinnitus, difficulty swallowing, and/or problems with eye movement.

17% of survey respondents reported having been diagnosed with Chiari II. When stratifying by age an inverse relationship was evident: 21–29% of people aged 25–35 said they had Chiari II, dropping to just 0–2% of people aged 66 and over. This is likely to be a reflection of improved antenatal screening techniques and consequent awareness of having the condition rather than an increase in rates of Chiari II in the younger generations. A number of respondents wrote that they did not know what Chiari II was, suggesting there is work to be done in raising awareness of the condition among people with spina bifida so that they know what symptoms to look out for.

Of the people who had Chiari II, 58% had symptoms that they had been told by a doctor were caused by the condition. Most people reported that their symptoms started between the ages of 31 and 40, though the median age was slightly younger at 26 (Figure 30).

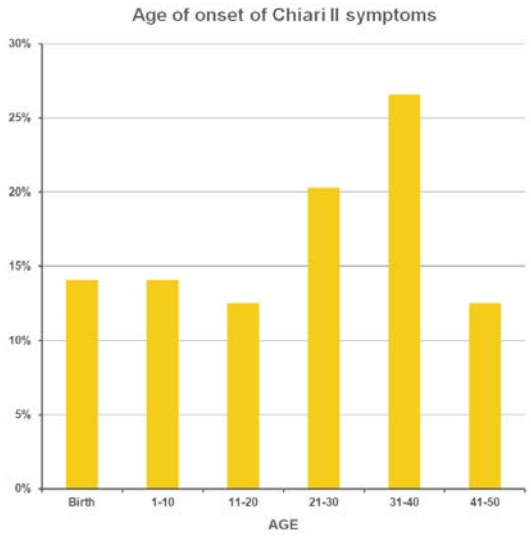


Figure 30: Age at which respondents began experiencing Chiari II symptoms

Surgeries that can be performed for Chiari II include shunting (to release intracranial pressure), detethering (if the cerebellum is being drawn downward by a tethered spinal cord), and decompression (where a portion of skull bone is removed to create extra space for the cerebellum). 26% of respondents with Chiari II said they'd had surgery for the condition. Most surgeries (23%) were performed between the ages of 21 and 30, and the median age was 21.

Section summary

Chiari II had been diagnosed in 17% of respondents. 58% of these had experienced symptoms, at a median age of 26. A quarter had been treated surgically (decompression, shunting, detethering), most often in their 20s. Healthcare providers of patients with open forms of spina bifida should consider Chiari II as a possibility if the patient is experiencing symptoms, and initiate a neurosurgery and imaging referral to investigate.

1.3.9 Tethered cord

Tethered cord (a.k.a. tethered spinal cord syndrome) is a condition caused by the spinal cord becoming anchored to tissues that limit the cord's movement within the spinal column during growth. The attachments can cause distension of the spinal cord, producing symptoms such as back pain, turning in of the feet (talipes), pins and needles in the lower body, thinning of the calf muscles, numbness in the feet, bladder problems (frequency, urgency, leaking, recurrent infections), bowel problems (constipation, soiling) and pain, weakness or fatigue during walking.

Tethered cord is frequently associated with spina bifida and symptoms often occur during periods of growth (e.g., puberty) but they can occur later in life as the spinal cord deteriorates after long-term tethering. 28% of survey respondents reported having been told they have a tethered cord.

The most common age for diagnosis was ≤10 years old (24%) and between 36 and 45 (22%); the median age of diagnosis was 34 (Figure 31).

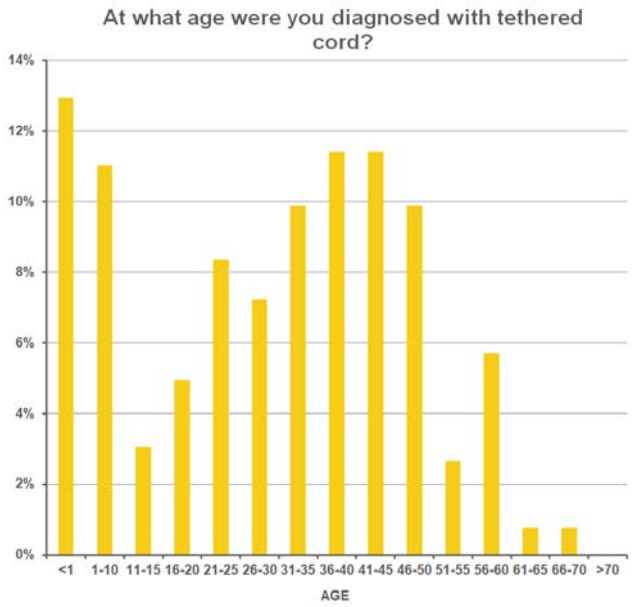


Figure 31: Age when respondents were diagnosed with tethered cord

The most commonly reported symptoms by those who had been diagnosed with tethered cord were pain (74%) and difficulty walking (71%). 49% had experienced pins and needles (Figure 32).

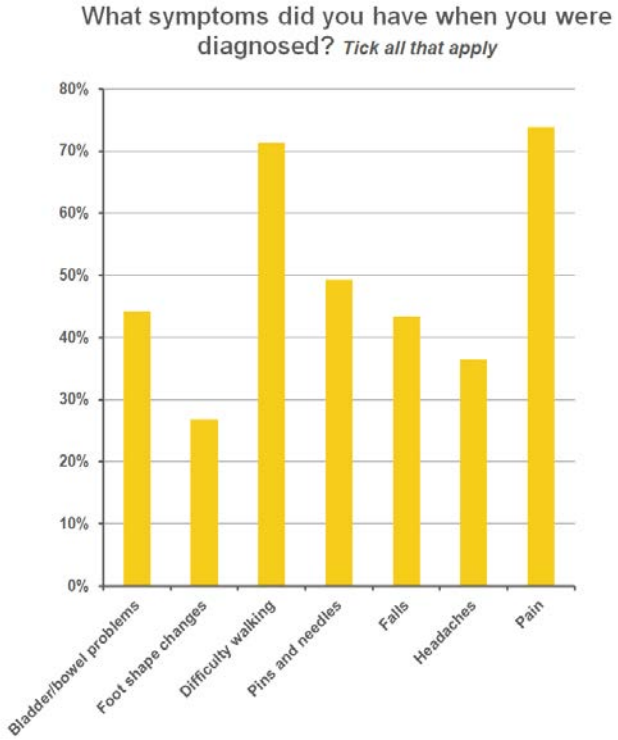


Figure 32: Symptoms of tethered cord experienced by respondents at the time of diagnosis

42% of respondents who'd had a tethered cord diagnosis had undergone detethering surgery, with the operation most commonly being performed in people aged 10 and under (27%), though the median age for surgery was 25 (Figure 33). 12% of those who gave ages for their surgeries reported having multiple detethering operations.

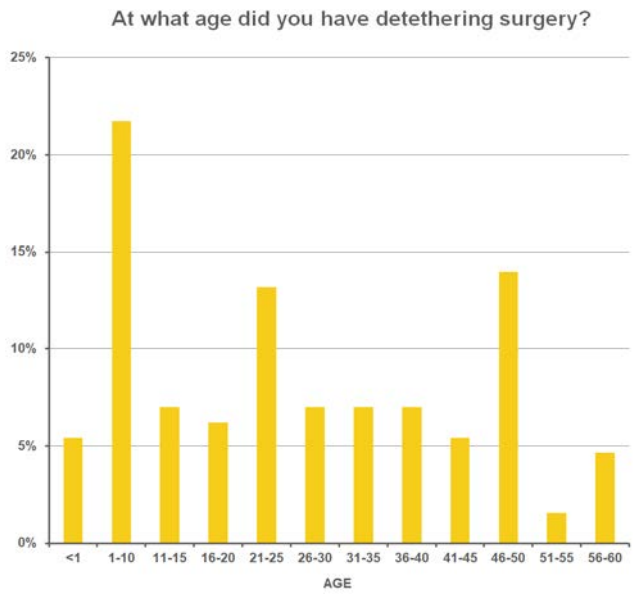


Figure 33: Age when respondents had detethering surgery

Section summary

Around 3 in 10 people with spina bifida had been diagnosed with tethered cord. Most had been diagnosed in childhood but many were also diagnosed in their early-middle years (36–45). When tethered cord is diagnosed in adults it's often a result of the spinal cord deteriorating after a lifetime of tethering. Imaging of the spinal cord at transition from paediatric services and careful follow up of people with tethered cord will detect deterioration at an early stage, and help prevent reducing mobility, worsening bladder/renal function, bowel function, and pain.

1.3.10 Kidney, bladder and bowels

Neurogenic bladder dysfunction (failure to store or empty urine) is a common feature of spina bifida. The bladder, the external sphincter, or both, may be affected to different degrees.

Even with clean intermittent catheterization, there is a significantly increased risk of urinary tract infections, and consequent scarring of the kidneys. Some people with spina bifida experience ureteric reflux, where urine from the bladder is squeezed back though the ureters to the kidneys.

This increases the risk of renal scarring and, without intervention, of chronic kidney disease. 37% of respondents had reduced kidney function (Figure 34).

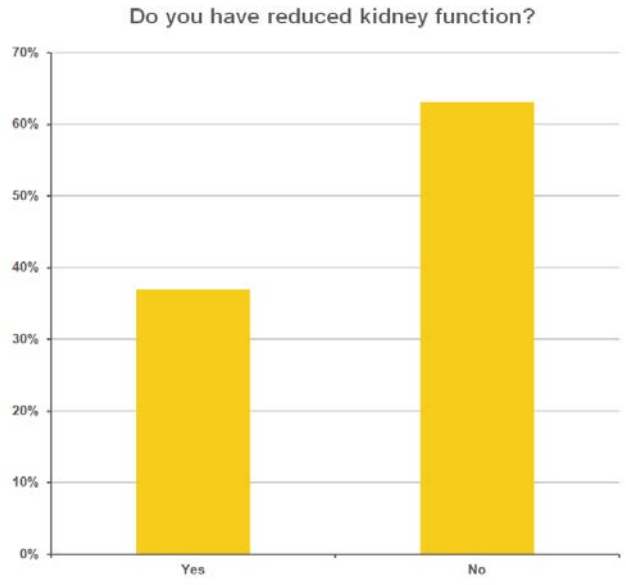


Figure 34: Percentage of respondents with reduced kidney function

The lifetime risk of developing kidney and bladder stones (urolithiasis) in the UK is 14% (Rukin et al., 2017). Urolithiasis is known to be associated with spina bifida (Veenboer et al., 2013) and accordingly we found that 26% of members reported having had kidney or bladder stones (Figure 35).

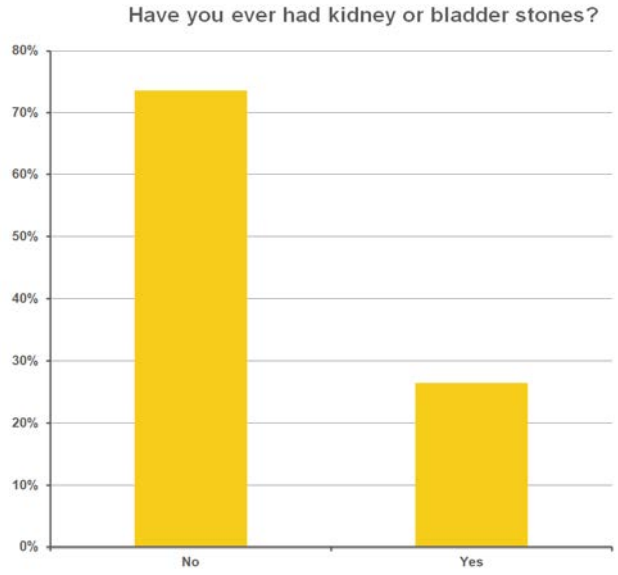


Figure 35: Percentage of respondents who had experienced kidney or bladder stones

Hypertension (high blood pressure) is a risk factor for a number of diseases – extra strain is placed on the heart and other organs, including the brain, kidneys and eyes. Having spina bifida increases the risk of hypertension, possibly due to renal problems (the kidneys are important regulators of blood pressure) or possibly due to increased obesity, reduced activity, and/or the association with sleep apnoea.

The prevalence of hypertension in people with spina bifida aged 16–59 years has previously been reported as 21%, though this was in the USA (Liptak et al., 2016). The prevalence of high blood pressure in the general adult population of England has been estimated at 26% (PHE, 2020).

We found that in our respondents with spina bifida the overall prevalence was higher, at 33% (Figure 36A). High blood pressure was broadly associated with increasing age, rising from 6% of 25–35-year-olds to a maximum of 51% in the 61–65 age group (Figure 36B). Over the age of 65 the prevalence of hypertension dropped slightly but not significantly.

Most people were diagnosed with hypertension between the ages of 36 and 50 (48%), and the median age for diagnosis was 40 years old.

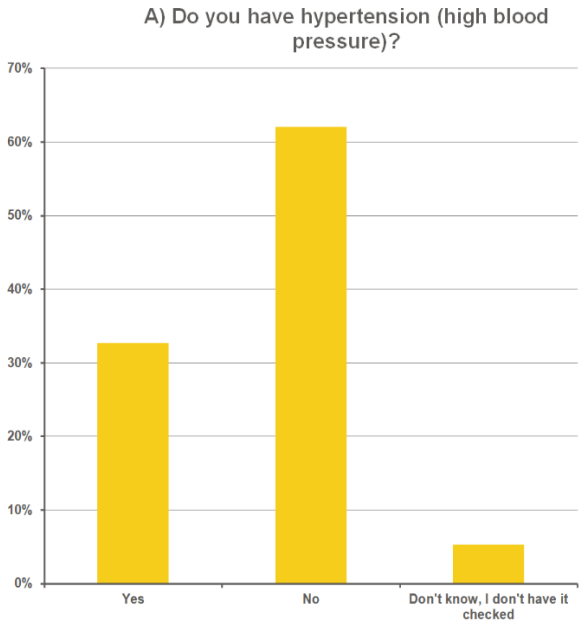
As is seen in the general population, men with spina bifida were at higher risk of hypertension than women: 43% compared with 26% (Figure 36C).

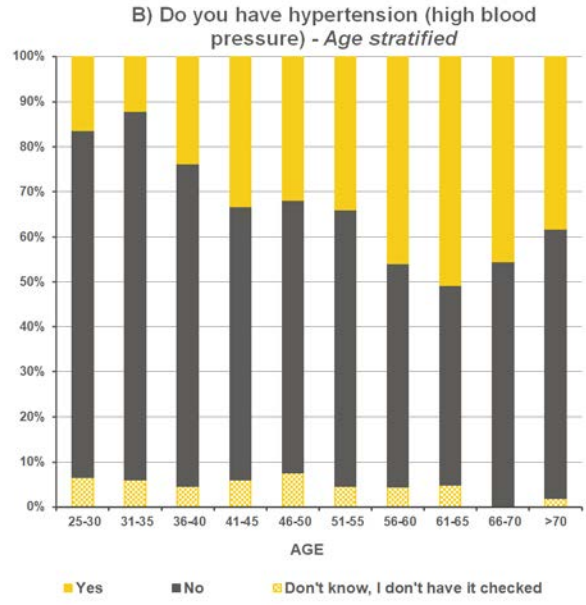
Interestingly, the sex-ratio for hypertension in the wider English population is 31% in men and 26% in women (PHE, 2017) which suggests that the increased overall prevalence of hypertension in spina bifida may be due largely to more males having raised blood pressure.

5% of respondents did not know/check whether they had high blood pressure, but the data showing increased prevalence of hypertension in people with spina bifida suggests this needs to change.

Hypertension is largely preventable through lifestyle modifications such as diet, exercise, and smoking cessation, and these factors can help to reduce blood pressure that has already become high.

There is a need for routine blood-pressure monitoring in the adult spina bifida population in the UK.





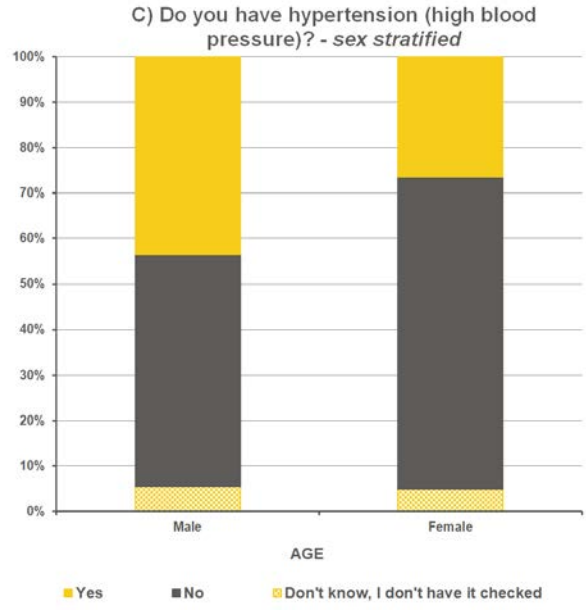


Figure 36: Hypertension prevalence in A) all respondents B) in different age groups and C) in males and females

Because bowel and anorectal function is controlled by spinal nerves, many adults with spina bifida have some level of bowel dysfunction, known as neurogenic bowel. This means that the majority of people with spina bifida may experience bowel leakage and some may also have constipation.

Bowel function can be managed in a range of ways but key strategies include adequate intake of fluid and fibre, a regular routine for bowel emptying, oral and/or anal medication, manual evacuation, irrigation (trans anal or via ACE or similar), and colostomy/ileostomy.

Bowel management is important for maintaining continence, for quality of life and long-term health. Our survey results show that the most common methods of bowel management among respondents were: naturally (55%), medication (24%), and manual evacuation (20%, Figure 48).

Most respondents reported opening their bowels every day (43%), or every 2–3 days (38%), but 12% reported opening them every 4–6 days, 4% every 7–9 days, and 2% every 10 days or less often (Figure 37).

There were significant differences between the sexes: males were more likely than females to open their bowels every day (50% compared with 40%), and females were more likely than males to open their bowels every 10 days or less often (3% compared with 0.6%).

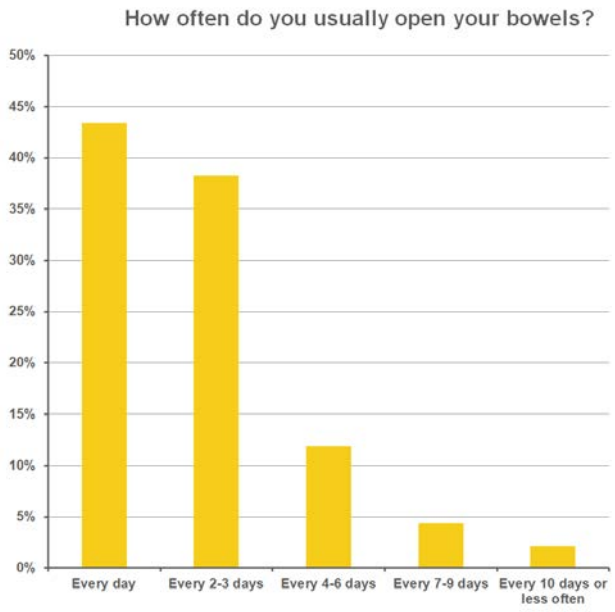


Figure 37: How often respondents opened their bowels

Soiling can have a significant impact on quality of life and mental health. 30% of respondents said they never experienced soiling and 29% soil once a month or less. 14% experience soiling once a week, 11% soil most days, and 16% experience intermittent explosive diarrhoea (Figure 38). The main difference that occurred with age was in those aged 61 and over; a greater proportion of respondents in this group (42–54%) reported never soiling, compared with those aged 60 and under (23–32%).

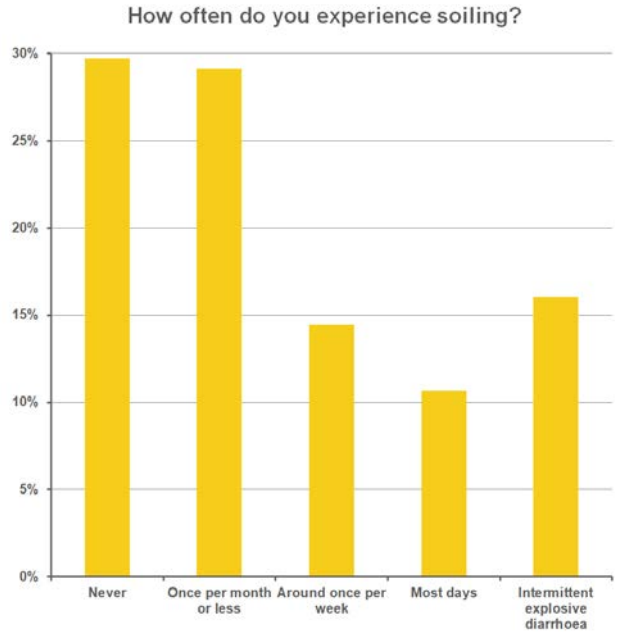


Figure 38: How often respondents experienced soiling

Some respondents shared their experiences with managing their bowels. This was not in response to a specific question, but the comments that were shared gave some insight into the lived experience of continence management. Constipation was mentioned often, and links between ageing, improper management, bowel problems, pain and diet were evident (Figure 39).

> *"from the age of about 30 I've had increasing worsening bowel function issues & I'm not seeing a bowel incontinence adviser"*

> *"I experience sever pain I relation to my poor bowel management - I experience in my head & face often"*

> *"stomach pain due to constipation. This pain radiates down my good leg across my back in my stomach"*

> *"I also often feel bloated due to my bowels so I don't eat properly"*

> *"Bowel problems - chronic constipation"*

> *"I use my Peristeen irrigation system 2 / 3 times a week with varying success. I've been seeing a bowel incontince adviser having many changes in medication - now know I've Slow Transit to start with"*

Figure 39: Unprompted comments about bowels and management

Section summary

Most people with all forms of spina bifida have a neuropathic bladder, strongly associated with bladder cancer, renal impairment, failure and premature death. Protecting the kidneys is in part dependent on maintaining a low-pressure, high-capacity bladder, and detecting changes at an early stage means treatment can be initiated promptly.

Bladder and renal ultrasounds, blood tests and blood pressure checks need to be done regularly to detect changes to the bladder, before irreversible renal damage begins. Recurrent urinary tract infections should be investigated and the underlying cause treated, to prevent renal scarring. Adults with spina bifida are more prone than people without to high blood pressure, owing to factors such as reduced mobility. That significant numbers were not having regular blood pressure checks alarmed us, and suggests a need for improved patient education and outreach.

Bowel incontinence is a major quality-of-life factor, as well as a risk factor for pressure sores. Most respondents soiled at least once per month. It is cited as a factor in leaving work, depression and anxiety.

Bladder and bowel function frequently deteriorate in adulthood and need monitoring throughout adulthood.

1.3.11 Mental health

Mental health refers to emotional, cognitive, and behavioural wellbeing. People with spina bifida may be at increased risk of depression and anxiety for a number of reasons: health issues causing distress, e.g., pain, illness, mobility/independence restrictions; cognitive effects of spina bifida and hydrocephalus can impact on how individuals process information, cope with emotions and respond to treatment; societal factors can impact on

wellbeing, e.g., public attitudes/prejudice, lack of access/adaptations, lack of autonomy, lack of access to suitable work/benefits/care.

Our survey results showed that 53% of respondents had experienced depression and 56% have had anxiety. Only 30% said they had no mental health problems (Figure 40A). These statistics are considerably higher than the average for adults in England, where 26% report ever having been diagnosed with at least one mental illness (Sally et al., 2015).

Only 12% of respondents reported that their mental health has improved over time, 49% said their mental health had stayed the same, and 39% said it had worsened (Figure 40B). 85% of people who'd had depression felt their mental health had stayed the same or worsened. 87% of people with anxiety felt their mental health had stayed the same or deteriorated.

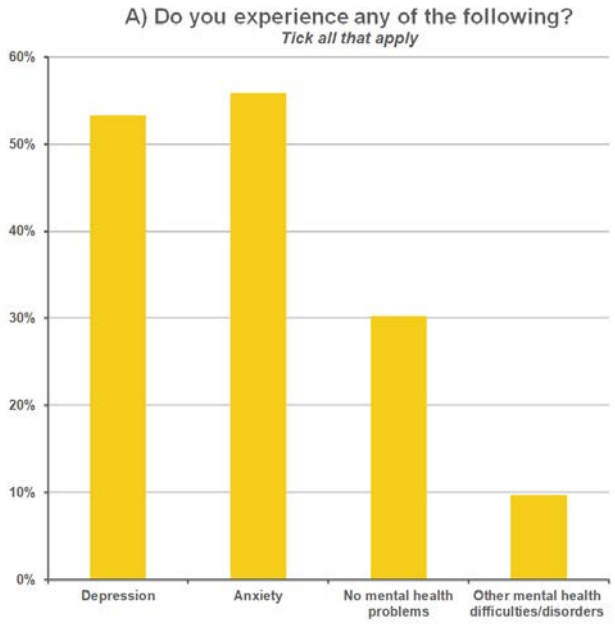
Having depression or anxiety was associated with many other issues. Respondents with these issues were:

- More likely to be unemployed (63% for anxiety and 63% for depression) compared with those with no mental health problems (56%). They were also less likely to be in full-time work: 11% for anxiety and 11% for depression, compared with 17% in people without mental health issues.
- More likely to report their mobility changing over time: depression (83%), anxiety (80%), no mental health problems (67%).
- More likely to have reported the movement in their legs reducing over time: depression (62%), anxiety (59%), no mental health problems (47%).
- More likely to have experienced deteriorating balance: depression (75%), anxiety (74%), no mental health problems (61%).
- More likely to experience shoulder pain and neck pain: depression (61% – shoulders, 57% – neck), anxiety (60% – shoulders, 56% – neck), no mental health problems (38% – shoulders, 34% – neck).
- More likely to have bone/joint disorders: depression (74%), anxiety (71%), no mental health problems (58%).
- More likely to have Chiari II: depression (21%), anxiety (22%), no mental health problems (10%).
- More likely to have tethered cord: depression (32%), anxiety (32%), no mental health problems (24%).
- More likely to experience some degree of soiling: depression (72%), anxiety (74%), no mental health problems (65%).
- More likely to have one or more symptoms of sleep apnoea:
- Tired every day: depression (65%), anxiety (63%), no mental health problems (26%).
- Snore loudly: depression (41%), anxiety (38%), no mental health problems (26%).
- Fall asleep during the day: depression (36%), anxiety (36%), no mental health problems (20%).
- More likely to experience pain 6–7 days a week: depression (57%), anxiety (53%), no mental health problems (38%), and less likely to experience pain rarely: depression (19%), anxiety (18%), no mental health problems (36%).
- More likely to report worsening pain over time: depression (68%), anxiety (66%), no mental health problems (52%).
- Less likely to feel their spina bifida was very well understood in hospital: depression (19%), anxiety (18%), no mental health problems (33%).

The relationship between mental and physical health was clear from the quantitative data, but also from comments shared by survey respondents. Declining health and lost independence were sources of particular upset for a number of people, as was feeling unsupported and overlooked by others, including medical professionals (Figure 41).

There is evidently a large unmet clinical need for mental health support in the adult population with spina bifida, and due to the large effect on quality of life and the known impact of mental health on physical health, this is something that needs to be addressed at scale.

To be maximally effective, any interventions introduced to tackle mental health issues in people with spina bifida would likely need to consider the range of cognitive differences in this population.



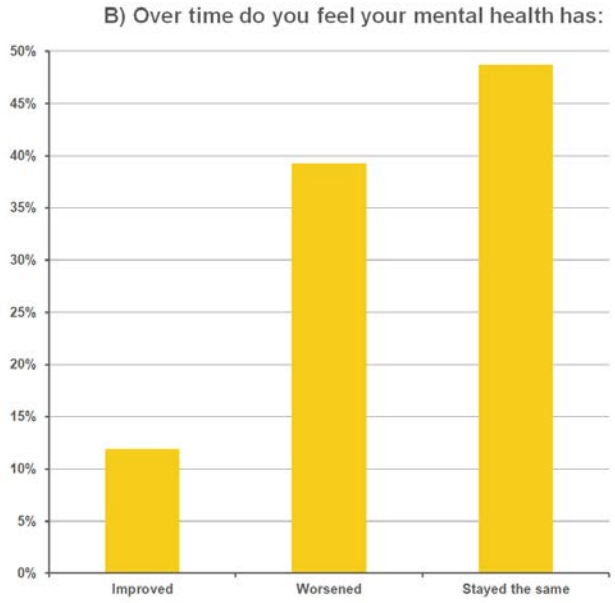


Figure 40: A) Respondents' experiences of mental health conditions and B) how their mental health has changed over time

> *"I wish that I had never been able to walk, and then I'd not have been unable to do my job, but also I've been overlooked, and left behind & ,let down, even by my family. Now there's nothing for me. I feel so low, so down and depressed to having feelings of suicide"*

> *"I have been used to getting around unaided all my life except for the last few years and it is making me feel frustrated and depressed"*

> *"I've had so many falls in my home but also outside, but the way people respond is what makes me feel scared to leave my home"*

> *"it is hard at times though to stay on top of thing with the bowel and bladders problems, life i mean its hard to live with at times"*

> *"I have a fear of hospitals due to spending 9 months in hospital when I was 3 years old"*

> *"surgery for self harm wounds - perineal abscess surgery - dehydration - 10 days for NG feeding due to anorexia when my weight fell below 6 stones"*

> *"Due to extensive damage to my shoulders and elbows (and more recently wrists, neck and spine) I have completely lost my independence. I am now reliant on my husband to help me with my personal care and household responsibilities. This has had a profound effect on my self-esteem and our relationship. I married the man I love, my husband is now my carer. I hate it!"*

> *"It's very frustrating not being able to walk the distances I used to and relying entirely on my partner"*

> *"in 2017. some of my occipital skull removed, no real explanation, no support from anyone. This has further impacted my mental health"*

Figure 41: Respondents describing their experiences with mental health issues

Section summary

Anxiety and depression each affect more than half of people with spina bifida and for around 9 in 10 their poor mental health has remained the same or worsened. Poor mental health was associated with significant problems e.g., unemployment, reduced mobility, increased pain. Whether poor mental health is a cause or effect of health issues, there is a substantial need for mental health support. Because of the profound link between pain and mental health, particularly anxiety, ensuring pain is effectively managed should also improve patients' quality of life.

1.3.12 Sleep apnoea

Obstructive sleep apnoea is a sleep disorder where breathing is interrupted by a narrowing of the airway caused by relaxation of the throat muscles and obstruction of the windpipe. Symptoms of sleep apnoea when asleep include: snoring, stopping breathing, or choking, and tossing and turning during the night. Daytime symptoms of sleep apnoea include: falling asleep during the day, not feeling refreshed after a night's sleep and instead feeling groggy and tired. Headaches are common, as are difficulties with memory and concentration. People with Chiari II are at increased risk of sleep apnoea, as are overweight/obese people with excess weight in the neck area. In the UK obstructive sleep apnoea is thought to affect 1.5 million people (approximately 3% of the adult population) (British Lung Foundation, 2015). Our survey data show that, relative to the general population, a high proportion of respondents had been diagnosed with sleep apnoea (10%), and that more males than females had been diagnosed with the sleep disorder (14% versus 8%). This is a similar trend (though with higher proportions) to that seen in the general UK population, where 4% of middled-aged males and 2% of females are thought to have the disorder.

67% of those diagnosed with sleep apnoea had been treated for the condition and 78% said it worked OK or worked well. Only 22% felt that it didn't work or weren't able to tolerate the treatment (Figure 42).

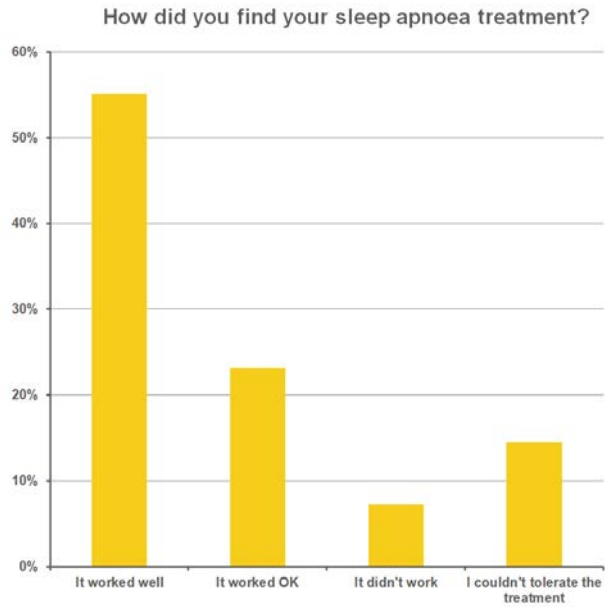


Figure 42: How well CPAP worked for respondents treated for sleep apnoea

In the undiagnosed population, 63% had at least one symptom and 51% had two or more (Figure 43): 50% felt tired most of the day, 34% snored loudly, and 30% fall asleep during the day without knowing why.

Because of the strong evidence that sleep apnoea is a significant risk factor for cardiovascular disease and mortality, it is vital that people with spina bifida and professionals involved in their health and social care are aware of the symptoms, to enable diagnosis and treatment.

It has been shown that people with sleep apnoea have higher blood pressure, and if the condition is untreated it is associated with arrhythmia, stroke and coronary heart disease (British Lung Foundation, 2015).

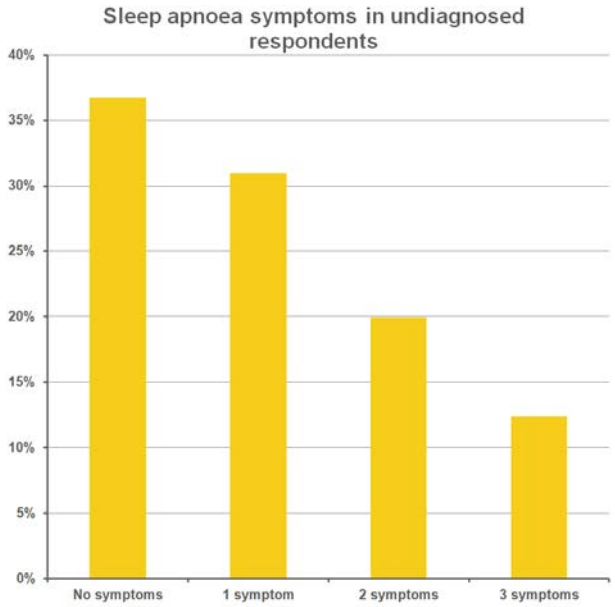


Figure 43: Prevalence of sleep apnoea symptoms in undiagnosed respondents

Section summary

People with spina bifida are at increased risk of sleep apnoea for a variety of reasons including Chiari II and an increased likelihood of being overweight. Sleep apnoea appears to be underdiagnosed in the spina bifida population, as around two thirds of respondents had at least one symptom. Patients with spina bifida and sleep apnoea generally respond well to treatment, so early diagnosis and treatment is key to prevent the long-term damage to the body associated with untreated sleep apnoea.

1.3.13 Pain

Long-term (chronic) pain is a debilitating condition where persistent pain signals impact on quality of life and daily functions. There may be a specific underlying cause such as injury, infection, inflammation, or general wear and tear.

Pain can also be caused by changes anywhere along the pain pathway i.e., nerves, spinal cord or brain. Because of the natural protective mechanisms that are activated when we experience pain, stress and anxiety are instinctively triggered as the body attempts to stimulate us into taking protective action. There is therefore a complex interplay between pain and mental health, particularly anxiety.

An estimated 30–50% of people in the UK have long-term (chronic) pain (Fayaz et al., 2016). Our survey revealed that 75% of respondents experience pain at least once a week and 47% are in pain 6–7 days a week.

Only 25% of respondents reported that they rarely experience pain. The proportion of people reporting experiencing pain 6–7 days a week generally increased with age, from 40–41% in the respondents aged 25–35, up to a maximum of 66% in people aged 61–65.

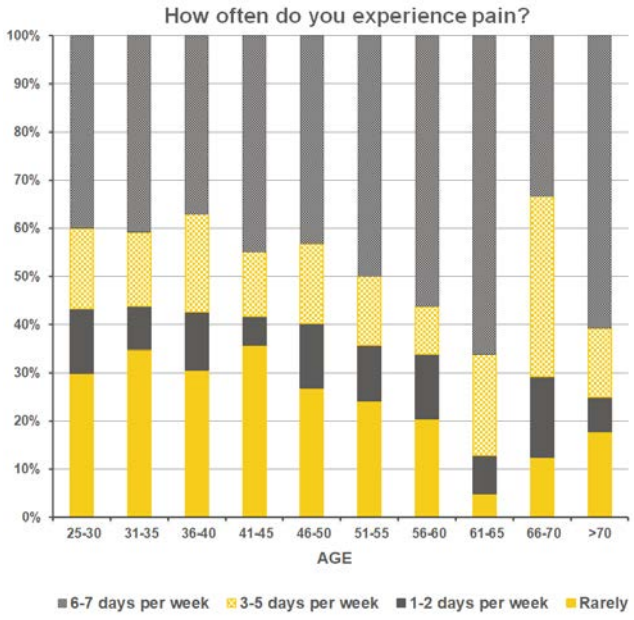


Figure 44: How often respondents experience pain

The inverse trend was also apparent: that the proportion of people rarely experiencing pain decreased with age, from 30–35% in those between 25 and 45 years old to 5–17% in the 61 and over population (Figure 44).

There was a small but significant difference in pain experienced between men and women: males were more likely to rarely experience pain (30% compared with 23% for females), and were less likely to experience pain 6–7 days per week (42% compared with 50%).

Due to the known relationship between pain and mental health, we examined the impact of these conditions on each other by data stratification: examining prevalence of mental health when stratifying by pain frequency, and examining the proportions of people experiencing different pain frequencies when stratifying by mental health condition (Figure 45).

The analyses showed that the more frequently people experienced pain, the more likely they were to experience mental health problems. In people who rarely had pain 43% had no mental health problems; in those who experienced pain 3–7 days a week 24% had no mental health problems.

The relationship was mirrored when stratifying by mental health issues experienced: people with no mental health issues were more likely to be rarely in pain (36% compared with 19% in people who'd had depression, and 18% in those who'd had anxiety).

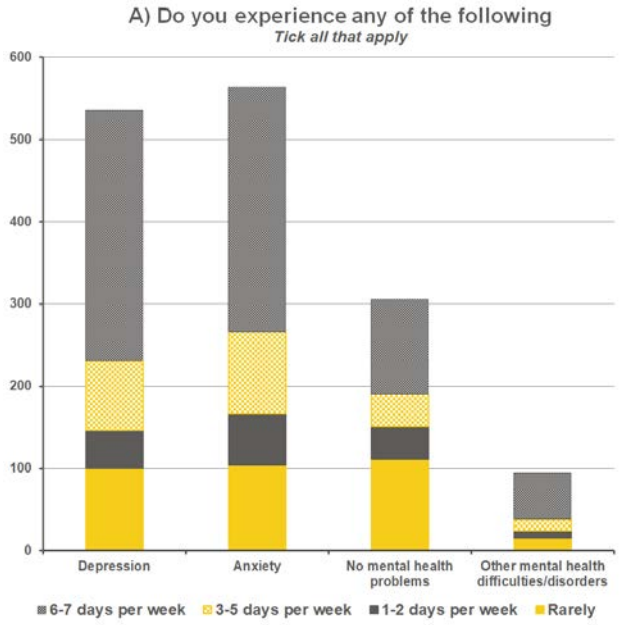


Figure 45 (above and overleaf): The relationship between mental health and pain

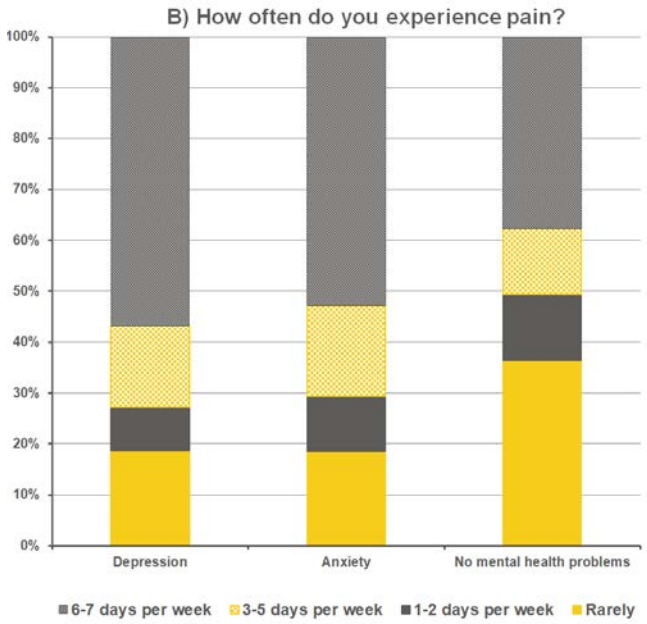


Figure 45 above: The relationship between mental health and pain

In many of the free-text boxes, respondents described the profound impact of pain on their quality of life (Figure 46).

> *"I am in pain of some sort most/every day. Some days are better than others"*

> *"The older I get the worse my pain becomes"*

> *"Pain is as much a limiting factor as function now. Years of walking badly have taken their toll on my joints"*

> *"I tend to get a lot of pain in my joints and I feel a lot of pain from my lesion area down to my legs. At times I experience a tingling sensation and this causes my bladder to be very irregular and incontinent"*

> *"In the last couple of years, I have had a lot of shoulder and hip pain which has reduced my ability to be as fit as I used to be"*

> *"Some days my legs are so painful my muscles ache. Cannot walk for long so don't go out"*

> *"As you get older the worse you feel"*

> *"Constant pain stops me from doing things"*

> *"As I have got older it has got worse. It has stopped me from working in school could not do my job. Some days I cannot wash my hair or dry it. Even getting dressed is a nightmare. I cannot go shopping by myself so I order on line now. I have no strength in upper body. Pain which causes headaches"*

Figure 46: Respondents' experiences of chronic pain and its impact

Section summary

Three quarters of people with spina bifida experience pain at least once a week – nearly half have pain 6–7 days a week. The frequency of pain increases with age and for most the pain also worsens. Pain significantly impacts on mental health and on independence. To improve patients' quality of life, pain management should be reviewed regularly. To prevent pain becoming chronic, early intervention is vital and prevention is better still where possible: helping patients to manage their condition and general health e.g., weight management and exercise/physiotherapy to protect joints, ensuring adequate fluid intake and regular vision checks to manage headaches.

1.3.14 Biological-sex specific questions

Menopause typically occurs from 45–55 years of age as oestrogen levels reduce in the body. The average age for a woman to reach menopause in the UK is 51, however ~1% experience premature menopause (menopause under 40 years old) (NHS, 2018).

Anecdotally, Shine members have suggested that females with spina bifida experience early menopause, so we asked female respondents whether they had been through the menopause and at what age they experienced their last period. When stratifying the results by age there was limited evidence to suggest that overall women with spina bifida go through the menopause significantly earlier than usual: only 1% of women ≤40 reported having gone through the menopause (Figure 47), in line with the rate in the UK population.

The median age for last menstrual period was 49, slightly under the UK average but well within

the typical range. 28% of women who had been through the menopause were prescribed hormone replacement therapy.

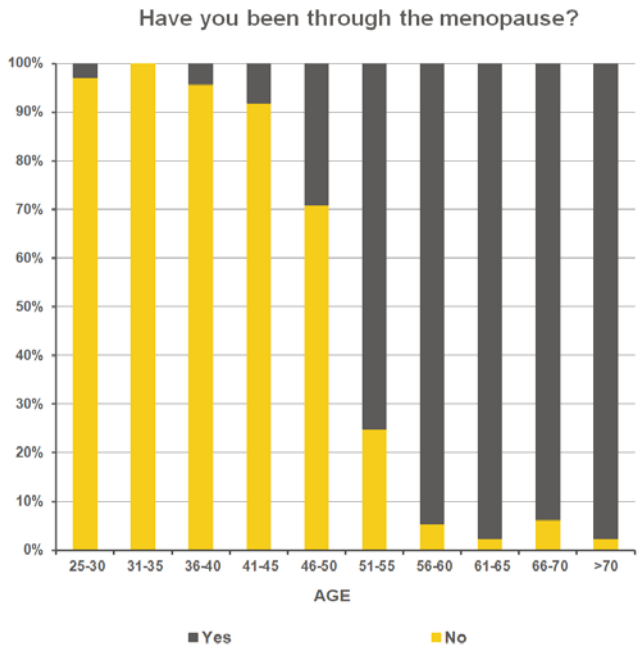


Figure 47: Ages of female respondents who reported having been through menopause

These results have health implications for females with spina bifida: no evidence of premature menopause in females, despite precocious or early puberty being associated with hydrocephalus, suggests some women with spina bifida will be exposed to oestrogen for more of their lives. A longer oestrogen window is a known risk factor for breast cancer, suggesting an increased importance for women with spina bifida to undergo breast screening. Participation in breast screening is known to be lower in females with disabilities (Floud et al., 2017), so it will be important to identify and address any barriers to screening in women with spina bifida. The use of hormone replacement therapy in women with spina bifida may also need to be considered in terms of possible risk of breast cancer versus known risk of osteoporosis.

An enlarged prostate (benign prostatic hypertrophy, BPH) is common in men over 50; approximately 40% will have evidence of enlargement in their prostate tissue and 33% will have symptoms (NICE, 2012). BPH prevalence increases with age, and by 80 years old 90% of males will have some degree of enlargement. The symptoms of the condition are mainly urinary, as an enlarged prostate can compress the urethra. Urethral narrowing slows the flow of urine during urination and can make it difficult to catheterise. We asked our male respondents whether they had been diagnosed with an enlarged prostate: 5% of all men under 50 reported having BPH, and 8% over 50 had the condition (Figure 48). There is no medical reason why men with spina bifida would be less predisposed to developing BPH and, due to their already compromised urinary system, there is a greater need for them to avoid further complications due to prostate enlargement. Greater awareness of the importance of prostate examination is needed among males with spina bifida and the professionals involved in their care.

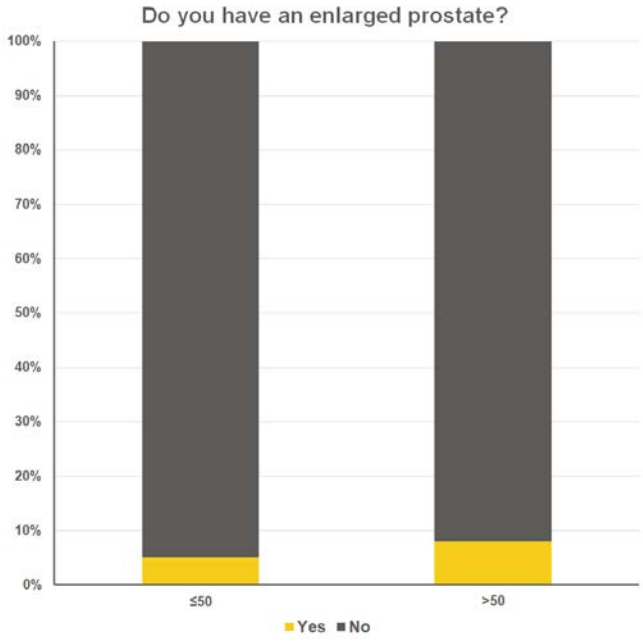


Figure 48: Prostate enlargement in males 50 and under, and over 50

Section summary

Due to increased prevalence of precocious puberty and menopause of typical onset, the oestrogen window that many women with spina bifida are exposed to will be greater. This is a risk factor for breast cancer, so breast screening in this population is essential.

An extraordinarily low number of men with spina bifida reported having prostate enlargement (only 8% of men over 50). There's no clinical basis for a reduced prevalence of BPH in spina bifida, so underdiagnosis is the likely cause. Prostate enlargement has significantly increased risks in spina bifida patients due to the effect of urethral compression on their ability to catheterise. Prostate examination is essential for men over 50 with spina bifida, and accommodations must be made to enable this: reminders to patients; adaptations to enable examination of wheelchair users.

1.3.15 Healthcare management and hospital admissions

As we have seen through the results already presented, people with spina bifida have complex health needs compared with the general population, creating significant challenges for healthcare.

Adults with cognitive impairment may find self-management of health and personal care particularly difficult due to the complexity and quantity of needs to be met. Significantly adding to the challenge is the relatively limited availability of specialist services and shortage of condition-specific expertise.

To get a picture of how spina bifida healthcare is being managed we asked in our survey about the types of healthcare people use and about their experiences.

74% of respondents have their healthcare needs met by seeing the GP when they are ill, and only 15% have regular checks with a multidisciplinary team (Figure 49A).

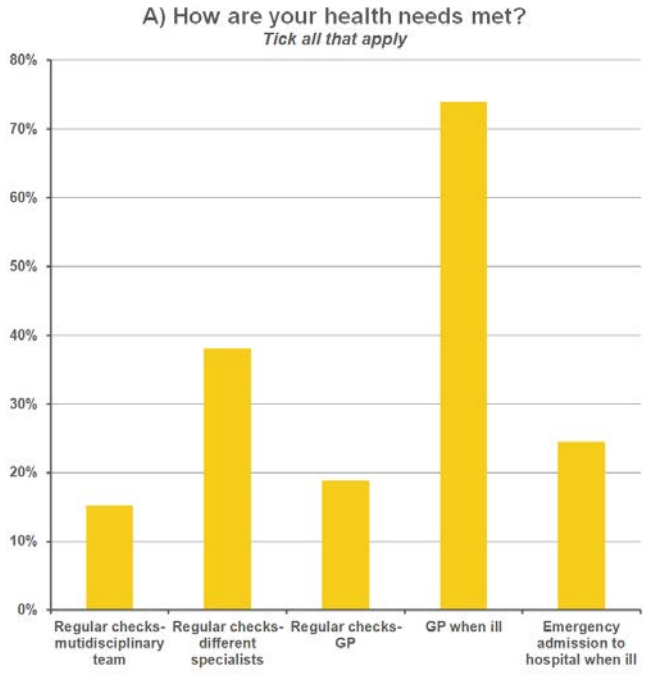


Figure 49: A) How respondents' health needs are met

Reactive care management (in response to illness/ emergency) was more commonly used than proactive checks: 58% compared with 42%; and non-specialist care (by GPs) was more commonly used than care by specialists (separately or part of a multidisciplinary team): 64% compared with 36% (Figure 49B).

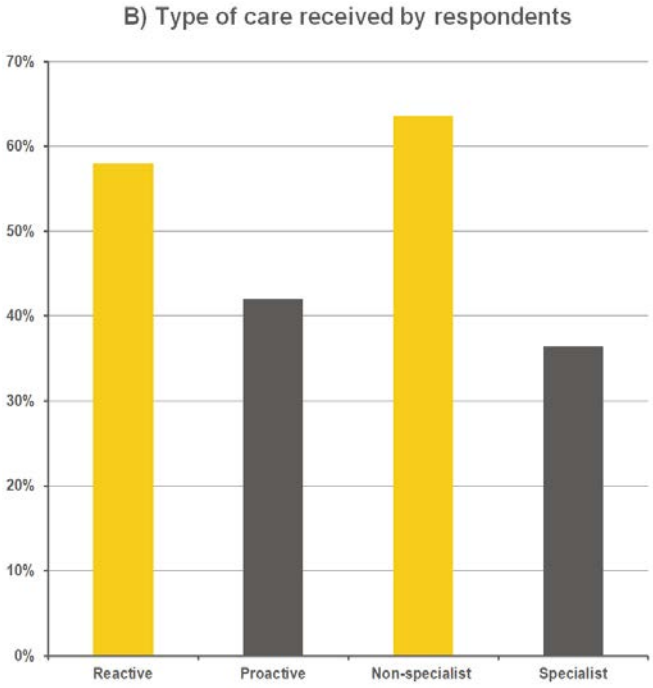


Figure 49: B) What proportions are proactive versus reactive care, and specialist versus non-specialist care

The most-seen specialists (Figure 50) were urologists (49%), followed by neurologists (21%), orthopaedic surgeons (19%), and neurosurgeons (19%). 25% of respondents reported seeing no specialists at all (Figure 50).

Of those who selected 'other', the most commonly seen specialists were podiatrists (13%) and gastroenterologist (11%).

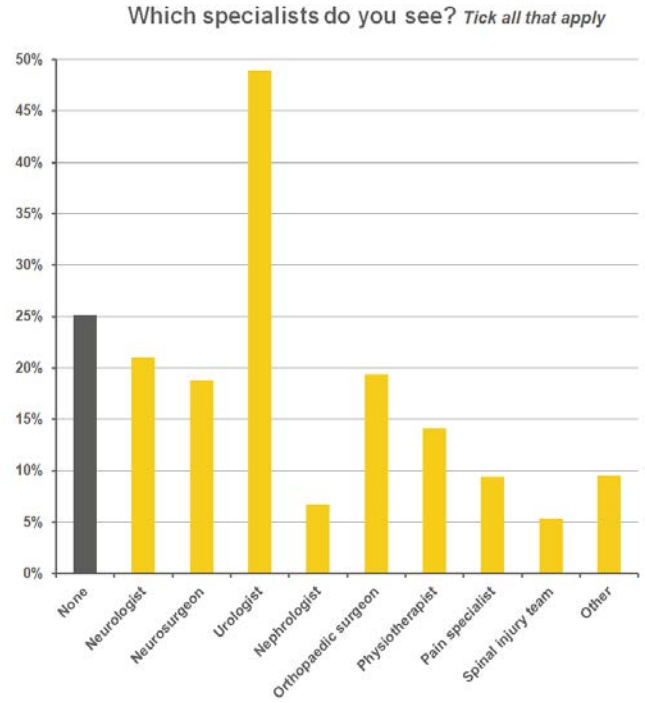


Figure 50: Specialists seen by respondents

Section summary

A quarter of people with spina bifida do not see any specialists and around 6 in 10 only receive reactive care rather than proactive/ regular support.

The health problems experienced by people with spina bifida as they age are complex and numerous. Many are interdependent and have the potential to be prevented or their impact minimised by early intervention, regular checks and reviews, and multidisciplinary approach to care.

1.4 Discussion

The changes in physical and mental health and cognition that typically come with getting older are generally well described in the wider population, but this is not the case for spina bifida.

This is largely because life-expectancy with the condition has only relatively recently been improved by medical advancements such as the shunt.

Ageing with a complex condition and meeting changing health needs may be made easier if some of those changes were better understood. This would enable targeted interventions to be found and implemented that minimise the impact of ageing on health and quality of life.

This study brought together a huge number and variety of experiences that have helped us to describe in unprecedented detail some of the key physical, mental, and social/lifestyle factors that change over a lifetime with spina bifida.

For the first time we have been able to describe quantitatively the lived experiences and challenges faced by people living with the condition in the UK. It is our hope that the data contained in this report will allow people with spina bifida to better understand their condition and to prepare for the future.

We also hope to use the report to better inform those involved personally or professionally in the care of people with spina bifida, to help them understand the challenges of ageing with the condition, and work with Shine to discover the most effective ways to help.

The next steps in the 1000 Voices Campaign will involve working with our members and with health and social care professionals to develop strategies for shaping the care and improving the lives of adults with spina bifida.

There is a clear need for ongoing monitoring of adults with spina bifida, and a proactive approach to healthcare management. This need was eloquently expressed by one of our respondents; it is an opinion we share, and that we now have the evidence to campaign for: **"Prevention rather than cure I believe would be a safer approach for those with Spina-bifida & Hydrocephalus."**

Shine
Spina bifida • Hydrocephalus
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1.5 Call to action

The needs of people living with spina bifida are complex and increase with age. Our report highlights numerous opportunities to reduce both the health and financial burdens of unplanned admission and ill health:

1. Use this report to adopt a proactive and coordinated approach to managing the healthcare of your patients/clients with spina bifida e.g., annual reviews of renal health, mobility, skin integrity, blood pressure.
2. Engage with your patients/clients with spina bifida to help improve understanding of their condition, and the importance of proactive health and condition management. Support younger people with transition and encourage independence.
3. Seek opportunities for multidisciplinary team working.
4. Join Shine as a professional member (membership is free and will allow us to update you about our work and campaigns).

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Appendix 1:

Estimating the number of responses needed

To calculate the required sample size for survey data to be accurately representative, the size of the total population is needed.

We estimate that there are between 11 and 14.5 thousand people living with spina bifida in England and Wales, and that 9.5 to 13 thousand of those are over 25. Using the most conservative estimate to ensure the most robust sample size, we took the upper estimate of 13 thousand.

We then needed to add the total for Northern Ireland, but unfortunately the data required to calculate an estimate is not available. However, we made a very rough estimate using the relative total population size of Northern Ireland (1.9 million) compared with England and Wales (59.5 million, combined). The population of Northern Ireland is 3.2% of the population of England and Wales, so assuming the birth prevalence and survival for spina bifida is the same then 3.2% of 13 thousand would be 418.

However, while we don't know the exact annual birth prevalence in Northern Ireland, we do know that it is higher than England and Wales, in part due to historical attitudes and legal restrictions on termination of pregnancy.

Again taking a conservative approach we added 2 thousand for Northern Ireland, for the purposes of determining the minimum size needed.

Our estimated population size for adults over 25 with spina bifida in England, Wales, and Northern Ireland was therefore 15 thousand.

Setting a 95% confidence interval and a 3% margin of error, the following formula was used to calculate the minimum sample size needed:

$$Sample\ size = \frac{\frac{z^2 \times (1-p)}{e^2}}{1 + \left(\frac{z^2 \times p(1-p)}{e^2 N}\right)}$$

N = total population (15,000)

z = Z-score for 95% confidence (1.96)

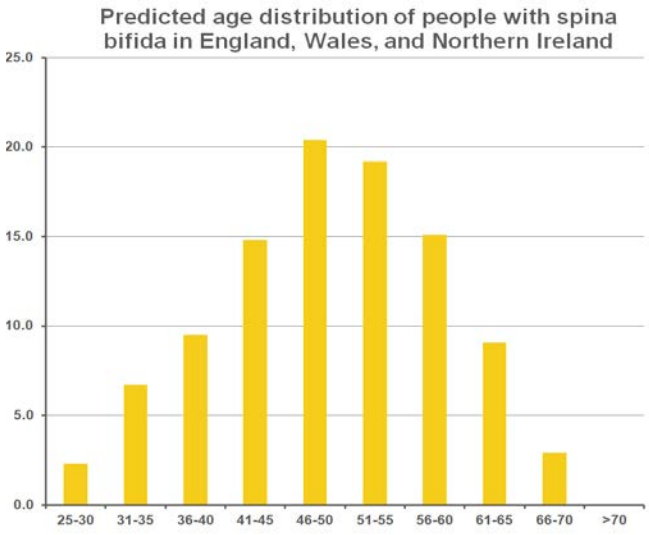
e = margin of error set at 3% (0.03)

This gave us a sample size of 997 which became our target and was the inspiration for our campaign name, 1000 Voices.

Appendix 2:

Estimated age distribution of people with spina bifida in England and Wales using spina bifida birth prevalence data and survival curves extrapolated from Oakeshott et al.

(Oakeshott et al., 2010) with 70 set as the maximum age.



SHINE Registered Office
42, Park Road, Peterborough, PE1 2UQ
Telephone 01733 555988
Email firstcontact@Shinecharity.org.uk
www.shinecharity.org.uk

Charity number 249338, Company number 00877990
Registered Office 42, Park Road, Peterborough, PE1 2UQ

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