

BMJ Open Psychosocial impact of the COVID-19 pandemic and shielding in adults and children with early-onset neuromuscular and neurological disorders and their families: a mixed-methods study

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To cite: Spurr L, Tan H-L, Wakeman R, *et al.* Psychosocial impact of the COVID-19 pandemic and shielding in adults and children with early-onset neuromuscular and neurological disorders and their families: a mixed-methods study. *BMJ Open* 2022;**12**:e055430. doi:10.1136/bmjopen-2021-055430

► Prepublication history and additional supplemental material for this paper are available online. To view these files, please visit the journal online (<http://dx.doi.org/10.1136/bmjopen-2021-055430>).

Received 13 July 2021

Accepted 23 February 2022



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ABSTRACT

Aim To describe and evaluate the psychosocial impact of the COVID-19 pandemic and measures to reduce the risk of transmission on patients with early-onset neuromuscular and neurological disorders (NMDs) and their families.

Design A mixed-methods study in which data were collected between 17 September 2020 and 31 December 2020 using a semi-structured telephone questionnaire developed specifically to meet research aims, and were analysed using quantitative methods and qualitative inductive thematic analysis.

Participants Forty questionnaires were completed by patients with NMDs (eg, muscular dystrophies, spinal muscular atrophy) or their parent. 70% (n=28) of patients were male, aged 2–48 years. 90% (n=36) were wheelchair users; 72.5% (n=29) required long-term non-invasive or tracheostomy ventilation.

Results Strict adherence to risk mitigation strategies, for example, shielding, were reported at the start of the pandemic. Over half continued some or all measures after official limitations were relaxed. 67.5% (n=27) reported changes to personal care assistance arrangements including temporary cessation of outside carers. Three themes were identified: (1) Concern regarding the health impact of COVID-19; (2) Perceptions of strategies to prevent SARS-CoV-2 transmission; (3) Psychological impact of the COVID-19 pandemic. The level and pervasiveness of frequently reported negative psychological effects, for example, anxiety and fear fluctuated, and were related to the perceived risk of COVID-19, concern about attending hospital, and perceived lack of access to intensive care management if severe COVID-19 infection occurred. Support, particularly from family and healthcare services, were considered to have positive psychosocial effects.

Conclusions Measures to reduce transmission of COVID-19 have greatly affected patients with NMDs and their families. For most, negative psychosocial impacts have and will continue to improve, but this may depend on the incidence of further pandemic waves. Consistent, up-to-date and accessible information on clinical outcomes and risk mitigation must be provided to support patients' physical and mental well-being.

Strengths and limitations of this study

- The identification of patterns of behaviour and specific areas of concern aims to inform guidance and interventions to support patients with neuromuscular and neurological disorders and their families, and may be generalisable to other vulnerable groups with complex disease and disabilities.
- Data has been presented as a description of trends through the pandemic to minimise effects of the study being conducted over several months during which the rates of COVID-19 and available guidance changed: this includes the subsequent availability of vaccinations for vulnerable patients which may have further reduced psychosocial burdens for many. In addition, perceptions of further lockdowns were not captured in this study and concerns may have changed. However, this study provides valuable insights into how to manage individuals in future pandemic waves.
- The study may be limited by selection bias with less representation of those at extremes of the clinical spectrum and those generally less concerned about COVID-19.
- Parents' perspectives are contextually different to patients' perspectives and their interpretations of psychosocial impact, including the mental and emotional well-being of their child may differ to that of the patient themselves.
- It is important to recognise the potential bias introduced by researchers involved in this study who are all clinicians working with patients from the study population, including some who participated; effects may include respondents avoiding providing negative statements particularly about healthcare services, however a sense of prior familiarity between researchers and patients/parents may have enabled more openness in participants' responses.

BACKGROUND

During the coronavirus 2019 (COVID-19) pandemic, strategies to minimise the transmission of severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) have included

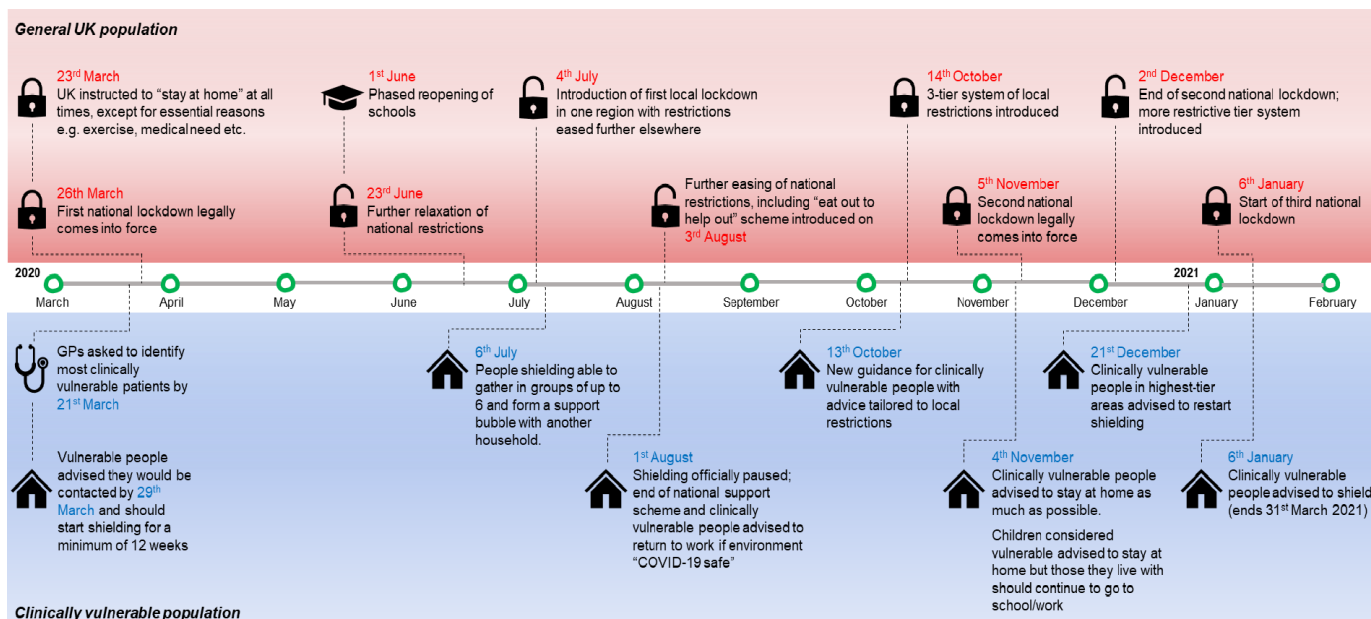


Figure 1 Timeline of restrictions in the UK for the general and clinically vulnerable populations. GPs, general practitioners.

targeted approaches to protect those at greatest risk: in the UK, people considered to be clinically extremely vulnerable were advised to ‘shield’ during peaks of infection¹ involving strict quarantine and avoidance of non-essential social contact (figure 1). Such strategies aimed to reduce the burden of excess deaths and the risk of overwhelming healthcare resources, but have been associated with negative impacts on physical and mental health^{2,3}: several previous studies reported high rates of stress, anxiety and depression at the start of the pandemic in general and disease-specific populations.^{4–6} In addition, changes made to healthcare services, predominantly through reducing face-to-face interaction may further impact on health outcomes.^{7,8}

Most patients with early-onset neuromuscular and neurological disorders (NMDs) were advised to shield during the pandemic: it was initially hypothesised that existing therapies, for example, corticosteroids and/or complications, for example, respiratory failure and cardiac disease^{9,10} would increase their risk of severe COVID-19. However, British, and European experience, supported by a limited body of evidence suggests that the clinical impact of COVID-19 in this population has been relatively low in terms of infection rate and severe outcomes.^{11–13} Reasons for this are likely to be multifactorial including the relative youth of patients with NMD, and the impact of minimising social contact. However, it remains poorly understood how those with NMDs approached and responded to risk mitigating strategies. We report the findings of a mixed-methods study investigating the psychosocial impact of the COVID-19 pandemic in adults and children with NMDs and their families. This information is intended to help care teams organise and adapt the support and services they provide during and beyond the pandemic.

METHODS

Design

This study used a parallel convergent mixed-methods design. Participants completed a semi-structured telephone questionnaire, which was specifically designed to meet our research aims. All questionnaires were analysed using quantitative methods and qualitative inductive thematic analysis. This enabled the description of patterns of behaviours and experiences during the COVID-19 pandemic, and interpretation of the associated and underpinning thoughts, motivations and psychosocial impact.

The questionnaire (see online supplemental materials) was developed in collaboration with members of the adult and paediatric home ventilation teams at the Royal Brompton Hospital (RBH), and a formal patient advisory group at RBH to ensure that the terminology and questions were appropriate and relevant to the target population. Included questions were informed by (1) the issues that had been raised in clinical and anecdotal experiences of the research team, and (2) emerging research in other patient cohorts, including those who were considered to be clinically extremely vulnerable and/or advised to shield during the pandemic. The questionnaire incorporated open and closed questions and comprised four sections:

- ▶ Personal and social circumstances prior to the COVID-19 pandemic.
- ▶ Experience of suspected or confirmed COVID-19 and access to acute healthcare.
- ▶ Changes to non-emergency medical care.
- ▶ Changes to personal and social circumstances.

All participants were asked all questions stated in the questionnaire. However, the order of questions could be varied by the researcher, for example, based on previous

responses, and participants were encouraged to elaborate on or clarify responses where they or the researcher felt initial responses were unclear, or further discussion would be beneficial to meet the study aims. Researchers conducting the questionnaires were clinicians with experience of managing patients with NMDs and as such were able and encouraged to use their knowledge and experience of the clinical and psychosocial context to clarify and explore responses in more depth.

Numerical, binary and brief descriptive responses were recorded in a questionnaire proforma by the researcher during questionnaire completion, and a complete audio recording of each questionnaire was subsequently transcribed verbatim for use in qualitative analysis.

Study population

Databases held at the study site were used to identify patients, who were assessed for eligibility by the direct care team. Adults or children with an early-onset NMD or their parent, who were able to provide informed consent were included. Parents completed questionnaires on behalf of those aged ≤ 18 years who remained under paediatric care, because patients were too young to answer independently and/or to prevent concern when discussing escalation of care and resuscitation which would not have been addressed with patients independently prior to transitioning to adult services. Patients' demographic data and medical history were obtained from electronic medical records. We adopted a purposive sampling strategy aiming to identify a variety of experiences and perspectives relating to our research aims; sample size was informed by (1) the number of questionnaires required to reach qualitative data saturation and (2) feasibility of recruitment from the available patient pool.

Statistical analysis

Demographic factors were summarised using mean (SD) for normally distributed continuous variables or median (IQR or entire range) for continuous variables that were not normally distributed. Questionnaire responses requiring numerical or binary (eg, 'yes/no') responses were reported as percentages of questionnaire respondents. Statistical analyses were performed using SPSS V.24.0.

Qualitative analysis

Transcriptions of all questionnaires were used in qualitative inductive thematic analysis performed based on the theoretical positions of Braun and Clarke.^{14 15} After familiarisation with the data set, LS and H-LT independently determined codes in a sample of six transcripts, chosen for their disparity. Codes were categorised and formalised, and after re-coding the initial transcripts, inter-coder similarity was assessed between LS and H-LT using NVivo.¹⁶ This process supported further rationalisation and confirmation of the final codebook (see online supplemental materials) and after discussion between LS, H-LT, RW, MC and AS, all remaining transcripts were

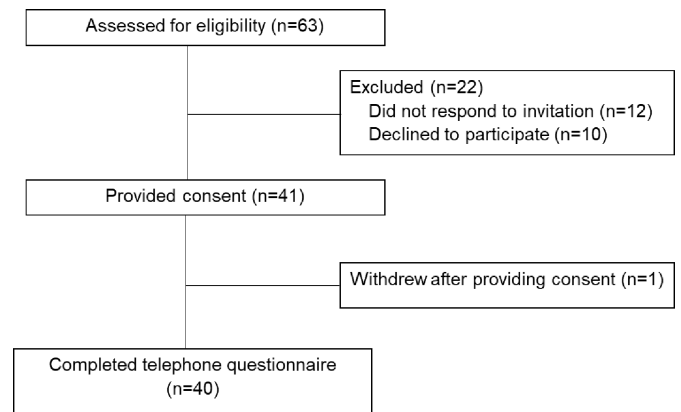


Figure 2 Flow diagram of participant recruitment.

coded independently by LS or H-LT. Candidate themes were determined by exploring coded data: the viability of these themes were reviewed by the coauthors and evaluated against the original data set before the final themes were defined.

Patient and public involvement

Patient participant advisory groups were consulted formally and informally during questionnaire development. In addition, the questionnaire was piloted among the research group and with patients and parents prior to use in the study. Participant validation was sought to provide additional confirmation of the accuracy of final themes, including by ZH, a patient representative and coauthor.

RESULTS

Forty participants completed the questionnaire between 17 September 2020 and 31 December 2020 (**figure 2**). As shown in **table 1**, patients were 70% male (n=28), aged 2–48 years and had early-onset NMDs (eg, muscular dystrophies, spinal muscular atrophy). Long-term non-invasive or tracheostomy ventilation was required by 72.5% (n=29) of the patients. Questionnaires were completed themselves by 75% of patients (n=30) (40% male (n=12), aged 17–48 years). Questionnaires were completed by parents of 10 patients ≤ 18 years who had not transitioned to adult services (100% male (n=10), aged 2–17 years). All patients resided in England and reported having received written advice to shield during the first wave of the pandemic.

Quantitative results

Changes to personal and social circumstances during the COVID-19 pandemic

All patients were reported to have shielded during the first lockdown period (**figure 3**); over half started shielding before being formally advised to do so.

Changes to personal care assistance arrangements were reported by 67.5% (n=27) to minimise interaction with non-household contacts: of these 59.2% (n=16) stopped all visits from external carers for between 2 weeks and

Table 1 Patient characteristics

Patient characteristic (n, %)	
Age group, years	
0–5	4 (10.0)
6–11	3 (7.5)
12–18	4 (10.0)
19–25	4 (10.0)
26–31	6 (15.0)
32–38	10 (25)
39–50	9 (22.5)
Gender	
Male	28 (70.0)
Female	12 (30.0)
NMD diagnosis	
SMA I	2 (5.0)
SMA II/III	13 (32.5)
CMD	10 (25.0)
DMD	5 (12.5)
Congenital myopathy	7 (17.5)
Other (eg, early-onset scoliosis, congenital myasthenic syndrome)	3 (7.5)
Respiratory support	
No routine respiratory support	11 (27.5)
Nocturnal NIV only	18 (45.0)
Nocturnal and intermittent daytime NIV	3 (7.5)
Continuous NIV use	6 (15.0)
Tracheostomy ventilated	2 (5.0)
Wheelchair user	
No	4 (10.0)
Yes	36 (90.0)
Requirements for personal assistance	
Independent	4 (10.0)
Requires assistance for some physical activities	8 (20.0)
Requires assistance for all physical activities	28 (70.0)
Residence pre-COVID-19 pandemic	
Own home	11 (27.5)
Family home	27 (67.5)
Supported or assisted living	2 (5.0)
Frequency of leaving residence pre-pandemic	
Every day	11 (27.5)
At least four times a week	14 (35.0)
Two to three times a week	4 (10.0)
Every one to two weeks	9 (22.5)
Less often than monthly	2 (5.0)
Work and education	
Full-time education	7 (17.5)

Continued

Table 1 Continued

Part-time education (including home-based and early years groups)	8 (20.0)
Full-time employment	12 (30.0)
Part-time employment	5 (12.5)
Part-time or ad hoc voluntary work	8 (20.0)

CMD, congenital muscular dystrophy; DMD, Duchenne muscular dystrophy; NIV, non-invasive ventilation; NMD, neuromuscular and neurological disorder; SMA, spinal muscular atrophy type I/II/III.

6 months, when all care was provided solely by family members and 25.9% (n=7) reduced the number of individual carers or the frequency of carers changing over. Changes were typically made from March 2020 and usual care assistance arrangements typically resumed between June and August 2020. Changes to patients' social and professional/academic activities were reported in 95.0% (n=38) and 77.5% (n=31), respectively. Financial difficulties were infrequently reported.

Impact of the pandemic on healthcare access: non-emergency medical care

Around 52% (n=21) reported cancellation or delay of one or more hospital appointment or admission since March 2020, including for potentially disease-modifying treatments, for example, nusinersen. Hospital appointment conducted as a remote consultation, either by telephone or video call, for at least one time was reported by 97.5% (n=39) and 32.5% (n=13) reported changes in communication or access to their general practitioner (GP), for example, the ability to order prescriptions online. Changes to community healthcare services were reported by 57.5% (n=23) which were provided remotely or postponed until staff and personal and protective equipment (PPE) were available. Around 27% (n=11) reported difficulties in accessing servicing for medical devices such as non-invasive ventilators or oxygen concentrators, or obtaining consumables, particularly antibacterial filters used in non-invasive ventilation (NIV) and mechanical insufflation–exsufflation devices.

Concerns related to COVID-19 and access to emergency healthcare during the pandemic

Seventy per cent (n=30) of participants reported being 'extremely' or 'very worried' about the risk of COVID-19 to the patient's health (figure 4) at the time of questionnaire completion.

No positive results from SARS-CoV-2 PCR testing were reported after testing began in March 2020. Around 32.5% (n=13) reported symptoms commonly associated with COVID-19 of whom only 38.4% (n=5) had a PCR test due to limited community testing, and 40.0% (n=16) underwent asymptomatic screening, for example, prior to hospital appointments. Of those reporting relevant symptoms, 53.8% (n=7) considered COVID-19 infection to be clinically probable, and the same proportion sought

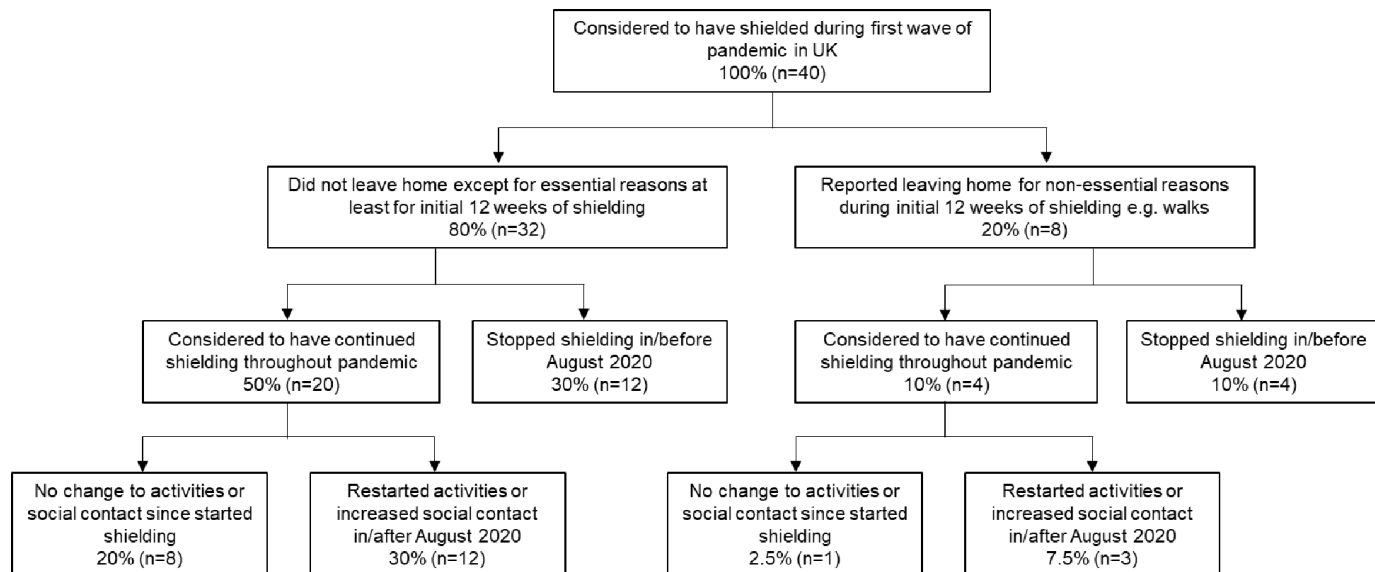


Figure 3 Reported patterns of shielding.

advice from their GP or National Health Service (NHS) 111 helpline; two patients attended the emergency department, but neither were subsequently found to have COVID-19.

Around 97% (n=39) of patients reported being for full escalation of care including resuscitation. Two patients had an advanced directive emphasising their wish for all treatment to be considered. Around 92% (n=37) reported they would wish intensive care unit (ICU) level care to be considered if they or their child developed severe COVID-19. However, of these 56.8% (n=21) were uncertain this wish would be respected by clinicians.

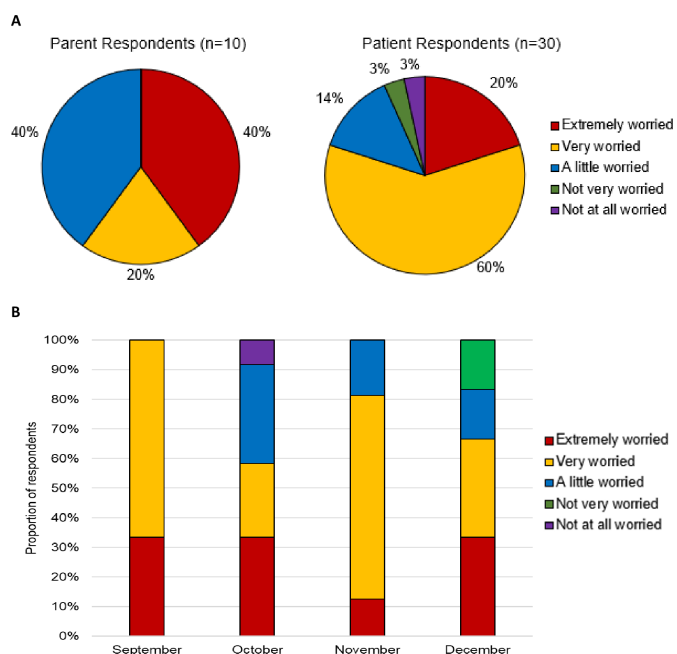


Figure 4 Reported level of worry about the risk of COVID-19 to the patient’s health at the time of questionnaire response (A) and as a proportion of participants by month of questionnaire (B).

Qualitative results

Three themes (table 2) were identified: (1) Concern regarding the health impact of COVID-19; (2) Perceptions of strategies to prevent SARS-CoV-2 transmission; (3) Psychological impact of the COVID-19 pandemic. Quotations reported below are followed in brackets by the respondents’ status as patient or parent, the patients’ gender, primary NMD condition and age group in years.

Theme 1: concern regarding the health impact of COVID-19

Risk of infection and disease severity

Concern about the risk of COVID-19 to patients’ health was associated with the perceived likelihood of severe or fatal disease. Perceptions were based on (a) previous experience of severe or prolonged illness due to respiratory infections and/or (b) vulnerability due to respiratory muscle weakness or failure:

...if a child cannot cough and clear their airways, any respiratory infection is going to be hard work. If your

Theme	Subtheme(s)
Concern regarding the health impact of COVID-19	<i>Risk of infection and disease severity</i> <i>Perceived accessibility and availability of acute care for suspected COVID-19</i> <i>Perceptions of prioritisation for ICU level care</i>
Perceptions of strategies to prevent SARS-CoV-2 transmission	<i>Shielding and minimising social contact</i> <i>Adaptations to home care arrangements</i> <i>Practical approaches to minimising COVID-19 risk</i> <i>Remote access to community and hospital care</i>
Psychological impact of the COVID-19 pandemic	<i>Psychological impact on patients and families</i> <i>Perceived positive outcomes from COVID-19 pandemic</i>

ICU, intensive care unit.

child has to have a PICU [Paediatric Intensive Care unit] or an HDU [High Dependency Unit] stay due to things like rhinovirus and been intubated because of a common cold, then COVID-19 obviously to me would pose the same risk. (Patient, male, spinal muscular atrophy (SMA) I, 0–5 years)

Because of my muscular dystrophy I'm classified as clinically extremely vulnerable. And I also sleep on a night ventilator because I have a compromised respiratory muscle system, so that concerns me quite a lot because COVID-19 is a disease that attacks your respiratory function quite quickly. (Patient, female, congenital muscular dystrophy (CMD), 32–38 years)

Uncertainty about the potential severity of COVID-19 was frequently reported, exacerbated by a lack of evidence specifically in NMDs:

...it seems a little bit like a lottery as to how [COVID-19] may affect individuals. Obviously I'm classed as clinically extremely vulnerable. But even in some of those cases, it seems a little bit hit and miss as to whether you would kind of see the full effect... (Patient, male, CMD, 32–38 years)

However, participants did not generally consider people with NMDs to be more likely to contract COVID-19 than others. Some considered COVID-19 to be of little risk to them, feeling they could entirely avoid infection, that is, through strict shielding.

Perceived accessibility and availability of acute care for suspected COVID-19

Reluctance to contact medical professionals in the event of suspected COVID-19 was not reported but unwillingness to attend hospital in the event of acute respiratory illness was common. This was because participants felt equipment and expertise was accessible at home:

...in some ways I'm quite lucky cos I've got all the equipment at home, so I've got the ventilator [and] cough machine, I've got all the things that they would do in the hospital anyway." (Patient, male, CMD, 32–38 years)

In addition, many thought their physical care needs would be less adequately addressed in hospital compared with at home, particularly if carers or parents were unable to stay due to restrictions on hospital visitors:

...I know that during the pandemic, they're not letting people stay with you. So that would put me in a very, very risky situation. (Patient, female, CMD, 39–50 years)

Several respondents reported being anxious about the risk of COVID-19 transmission in hospital, particularly if they were in close proximity to other patients. Others reported concerns based on pre-pandemic experiences about the ability of hospitals, particularly those without specialist expertise, to manage the complex needs of patients with

NMD; some considered that a personalised 'crib sheet' they or treating clinicians could follow would be useful:

I think it would be useful... to have a guide as to if something did happen, in terms of like my lungs... what the best tips are... (Patient, female, CMD, 32–38 years)

Perceptions of prioritisation for ICU level care

Concerns about ICU care not being considered or available were based on (a) the perceived lack of ICU capacity particularly during peaks of COVID-19 infections, and (b) perceptions that people with disabilities and underlying health conditions were considered lower priority:

...there was quite an outcry from the disabled community that hospitals were deciding not to put certain people, certain disabilities on a ventilator... that was quite a worry. (Patient, female, SMA II/III, 39–50 years)

...it's almost like they talk about like the death of a vulnerable person is just a bit inevitable... like they're the people that do die. (Patient, female, SMA II/III, 26–31)

Some adult participants had been advised by their specialist teams during the first wave of the pandemic that they would not be considered for ICU level care: some considered this an understandable stance aiming to prioritise those most likely to survive, but for most it increased anxiety, stress and anger:

...even in the best of times I would not go to hospital, but seeing this written, saying to you "if you get it, don't go to hospital", and it's the hospital saying that to you, that really brings your level of trust to zero. (Patient, female, CMD, 39–50)

There was a perceived lack of clarity in the criteria or process of prioritising patients for ICU level care. Some reported an awareness of the Clinical Frailty Scale (CFS) and were concerned that they would automatically be excluded or considered lower priority by virtue of their physical limitations. There was however doubt whether exclusions based on underlying conditions would be upheld in reality:

I would have hoped that if in the most severe cases, anybody that is suffering from COVID-19, regardless if you're classified as highly vulnerable... if you were suffering severe symptoms, I would have hoped they would have had space in ICU... (Patient, male, congenital myopathy, 19–25 years)

Theme 2: perceptions of strategies to prevent SARS-CoV-2 transmission

Shielding and minimising social contact

Initially patients and families aimed to avoid infection at all costs; patients' homes were considered places of safety where social contact could be controlled. In addition, virtually all patients reported having received adequate support to shield, for example, from friends and family,

employers, schools, healthcare services, and/or government support schemes.

...I haven't really left home since it all started. I get my shopping done online, the other half still works from home, so we're keeping away from people. (Patient, female, congenital myopathy, 32–38 years)

However a lack of clarity in confirming that people with NMDs were considered to be clinically extremely vulnerable to COVID-19 meant some only received formal advice to shield several weeks into the first lockdown, and several felt frustrated or let down by the lack of guidance provided by both the government and their clinical care teams:

No one really knows if it's the GP that's supposed to... or whether it's the clinical teams that are meant to decide. No one actually really knows who is meant to be telling them if their child is extremely vulnerable. (Parent, male, SMA I, 0–5 years)

Patterns of behaviour encompassed in 'shielding' varied greatly between respondents; deciding on when and how strictly to restrict activities and social contact were based on (1) national and local advice, (2) availability of activities, (3) perception of infection risk and/or (4) perceived impact on physical or mental well-being.

I was watching how the cases were starting going up again, and I thought that's it, I'm going back in. (Patient, female, CMD, 19–25 years)

Fundamentally shielding could be described as a state of mind informing acceptability of activities and social contact:

...I've just carried on doing what I have been doing since March [2020] going out when I really need to or when I want to, but avoiding contact with people... I'm still shielding, in my head, but I'm doing it according to what I feel intuitively is the right thing. (Patient, male, SMA II/III, 39–50)

Most patients continued shielding in some form outside periods where it was officially advised; household members generally returned to school or work, supported by strategies to minimise contact with others. However, some felt unsupported where they felt the household continuing or returning to shielding was in the patient's best interests:

...it's only guidance... it's not law... all I'm asking you to say, is that [I am] in a specific situation that requires a different sort of approach.... (Patient, male, SMA II/III, 26–31 years)

Many planned to continue shielding to some degree until vaccines were available, and often beyond to evaluate effectiveness:

...hopefully the vaccine is kind of successful and that's the end of it, but the reality of it probably isn't... we

can't be shielded forever... (Patient, female, CMD, 32–38 years)

Generally, participants were reassured by emerging evidence that young people and children were at less risk of severe COVID-19, but others remained uncertain:

...so obviously now a lot of children have been taken off the shielding list [and] he's been going to school. And every day I question myself, am I doing the right thing? (Parent, male, SMA II/III, 6–11)

Adaptations to home care arrangements

For patients requiring physical care assistance and support, complete isolation from others was not possible during the pandemic. Temporary cessation or reduction in external care provision was often undertaken to reduce the risk of COVID-19 transmission, and involved consideration of the risk posed by individual carers, for example, where they also provided care to other clients or in care homes:

I've been isolating, shielding, all this time but actually I can't truly isolate or shield, because I have full time care, so I'm very aware that every time any one of my six or seven people come to the door, they could be bringing it in. (Patient, male, SMA II/III, 39–50 years)

Such measures increased anxiety about the fragility of care arrangements:

If we were to get ill how do we look after him regardless of whether he gets ill or not? (Parent, male, SMA II/III, 12–18)

Pre-pandemic care arrangements resumed either when provision of all care by family became physically or emotionally unsustainable, rates of COVID-19 had reduced or protective measures, for example, PPE were available:

I think [the carers returned] when we realised there's PPE to help reduce the transmission, and it then got too much for my parents as well. 'Cos it was round the clock care so, it was too much to keep up with really. (Patient, female, CMD, 19–25 years)

It was also important for patients and families to trust carers to minimise their own infection risk and report potential exposure to COVID-19. However, this heightened concerns about establishing trust in new carers which exacerbated long-standing difficulties in recruitment:

...you know if somebody new was coming in I obviously couldn't be totally sure that they had done all the right things. (Patient, male, Duchenne muscular dystrophy (DMD), 26–31)

Practical approaches to minimising COVID-19 risk

Practical approaches to reduce the risk of transmission predominantly related to visitors, for example,

non-household family members, external carers. Strategies included increased hand hygiene and PPE. The type and form of PPE varied based on (1) availability of equipment, (2) available guidance, (3) perceived infection risk and (4) the presence of potentially aerosol generating procedures.

I mean social services have given us PPE that we're using... we're just mainly using gloves... My carers are basically taking precautions. (Patient, male, SMA II/III, 39–50 years)

Participants considered the risks of potential transmission of COVID-19 between carers and themselves or their child, particularly where carers worked with other clients. In addition, there was particular concern where the patient or family directly employed carers and were therefore responsible for their safety at work. Hesitancy of carers to use PPE, for example, due to discomfort was infrequently reported. When well, some adult patients did not feel PPE was required in their homes, believing it would be of limited benefit in preventing transmission or would impact on the quality of support or interaction:

I said that I'd prefer that [carers] didn't wear masks and aprons unless they absolutely felt they had to... Just purely because it felt very uncomfortable... very clinical. (Patient, male, DMD, 26–31)

...once they're indoors and once they're actually with me, the care is so intimate like, they're taking me to the toilet, taking me to shower, putting my mask on, actually PPE isn't really going to do anything. We're in the same space, breathing the same air for the whole week. (Patient, female, SMA II/III, 26–31 years)

Many participants reported trying to maintain social distancing with others, including carers, whenever possible. A minority reported ongoing concern about the potential for fomite spread of SARS-CoV-2 and had established extensive cleaning routines, for example, of groceries, the home, which increased physical and psychological burden:

...whenever we get something, I always ask my mum to wipe it and she gets annoyed at times, obviously, because I'm constantly saying it again. (Patient, male, congenital myopathy, 32–38 years)

Remote access to community and hospital care

Generally remote (virtual) access to healthcare services was viewed positively by participants as an interim measure to maintain contact with services, particularly for discussion-based consultations, and for patients who had maintained clinical stability over preceding months or years:

The thing is my condition is fairly stable, so my annual check-up is very quick usually. It's like me going in and saying "yep everything's ok" and then leaving

again. So, it was better to do it online. (Patient, male, SMA II/III, 39–50 years)

There was significantly less support for remote consultations where physical examination, procedures or diagnostic tests were deemed essential:

Basically he has been growing so quickly and his eyes, ears and teeth are constantly changing. And obviously those are the things we would like the medical person to have a look at, rather than just discussing over the phone. (Parent, male, congenital myopathy, 0–5)

Some participants preferred remote consultations reporting that they increased convenience and comfort, and mitigated logistical problems, for example, parking difficulties, limited wheelchair accessibility.:

...I found [remote clinic] a lot better, 'cos obviously, when you're talking from your own house, you're a lot more relaxed than when you are in hospital surrounded by everybody. So I mean you have to step into the room, and then you wonder if the room's a bit too crowded, or if you can fit through the door with the wheelchair. (Patient, male, SMA II/III, 32–38 years)

However, it was the preference of most participants that some, if not most, appointments return to being in person to facilitate communication and/or the performance of tests and procedures:

I think with the sleep clinic it's important to hopefully get back to coming down and having an appointment face-to-face because it means we can do the blood gas test and it gives you a view of whether the NIV's working properly. That's obviously something that we can't do via teleconference. (Patient, male, CMD, 32–38 years)

Theme 3: psychological impact of the COVID-19 pandemic Psychological impact on patients and families

Anxiety, fear and worry were the most frequently reported emotions during the COVID-19 pandemic (figure 5) although the level and pervasiveness of these fluctuated:

Honestly, it has varied across the past 8 months... I think in the beginning I was very worried, I was very anxious... (Patient, female, CMD, 39–50 years)

Primarily, negative emotions were related to (1) uncertainty, including about the potential impact of COVID-19 on the patient's health, (2) conflicting or changing guidance or advice and (3) the potential duration for which restrictions would be required. Depression and low mood were less frequently reported but were more common in those with previous mental health problems:

I'd say I've become a lot more depressed and I suffer from depression, so that adds on top of it. (Patient, male, SMA II/III, 32–38 years)



Figure 5 Responses to the question ‘How have you felt during the COVID-19 pandemic?’ by (A) patient respondents and (B) parent respondents.

Some reported negative psychological effects on the family when they were shielding or providing all patient care:

Some days I’m okay, some days I find it hard... when it was just me and the kids for 4, 5 months... all day and no adult conversation at all, that was quite difficult. (Parent, male, SMA I, 0–5 years)

For some, these feelings were exacerbated by seeing the general population being able to resume more activities as restrictions were lifted:

I think when the restrictions eased, and you keep seeing people on social media, boasting "oh, we’re going there", it does kind of make you feel left out. Like the world is moving and you’re just in the same place. (Patient, female, SMA II/III 39–50 years)

Others were more comfortable being at home as they felt protected or could continue activities they enjoyed:

...I feel safe in my house, where if I went outside I don’t feel safe at all. (Patient, female, early-onset scoliosis, 39–50 years)

Most patients missed social interaction with non-household contacts. However feelings of loneliness and isolation were uncommon due to mitigating factors, for example, keeping busy with work or schooling, maintaining contact remotely and interacting with carers and household members:

I spent the lockdown with my parents who I have a pretty good relationship with. So I wasn’t on my own. I really feel sorry for people who or were living alone

during the lockdown because how they managed to continue and keep their heads above water, and being completely alone, I have no idea. (Patient, male, CMD, 32–38 years)

Negative emotions generally improved during the pandemic, either due to (1) acceptance or adaptation to the situation, (2) better understanding of COVID-19 or (3) the resumption of some social contact or activities:

I have periods of stability where I feel quite safe. I think the uncertainty at the beginning did make me very anxious... I feel like I’ve got good coping skills on the whole so if I was anxious it wouldn’t last for days. I have gone up and down a lot, but I think that goes for everybody... (Patient, female, CMD, 39–50 years)

However, for some, negative emotions had been superseded by boredom, frustration and apathy. Particularly for those who shielded more strictly, there was anxiety about returning to normal activities and interactions:

...being at home, the [outside] world becomes a bit more of a scary place. And so I don’t know how I’m going to fit back in to doing things that I used to do. (Patient, female, SMA II/III, 26–31 years)

Several participants felt hopeful for the future or psychologically resilient and able to use coping strategies effectively. Some had taken proactive steps to improve their mental well-being, including seeking support from psychological medicine professionals:

I found that due to the sense of loneliness I was a little bit more sad, and maybe thinking in a negative way. And that’s when I asked for psychological support, and I’m now having like some sessions on a weekly basis with a counsellor. (Patient, female, CMD, 26–31 years)

Perceived positive outcomes from COVID-19 pandemic

Although predominantly a negative experience for most, almost all participants identified at least one positive aspect to come out of the COVID-19 pandemic. These were mostly related to (1) changes in personal circumstances, for example, spending quality time with family, or (2) impacts considered positive for wider society:

I think my relationships with family have become stronger... because we’ve been put into isolation, lockdown with our families for a really long time... (Patient, female, congenital myasthenic syndrome, 32–38 years)

...I think it’s brought to the surface a lot of inequality, which lots of people knew was already there. But now it’s a bit unavoidable. I think it’s come in to more of a public conversation and I hope that in terms of disability that continues... (Patient, female, SMA II/III, 26–31 years)

Of particular benefit was improved accessibility, as more activities and events were available online. Several participants reported that the pandemic had led them to re-evaluate their lifestyle and priorities, prompting the uptake of new skills, ways of working and increased focus on health and well-being:

...there were so many events that I used to go to that weren't accessible, or, [hadn't] got wheelchair space, and I had to miss out. Now they are online... equality was created, but at a price. (Patient, female, CMD, 39–50 years)

DISCUSSION

Principle findings

Anxieties about potentially severe clinical outcomes of COVID-19 were the main drivers of significant changes to lifestyle and healthcare access made by patients with NMDs and their families. The initial desire to avoid infection 'at all costs' resulted in severe limitations on contact with non-household members: this included formal and informal carers, which often resulted in increased physical and emotional burdens on households which were not sustainable indefinitely. Patterns of behaviour encompassed in 'shielding' were more variable as the pandemic progressed. Patients and families were required to make frequent and often numerous assessments of the risks, that is, of COVID-19 transmission, and benefits, for example, to their physical and psychological health to decide how and when to resume more normal activities. Many expressed that they felt unsupported in making these decisions which contributed to feelings of uncertainty and anxiety. However, negative emotions including fear and anxiety fluctuated throughout the pandemic, and overall decreased over time: many respondents were reassured by emerging evidence suggesting COVID-19 in NMDs and children may be less severe than predicted.^{13 17} In addition, there was hope that successful vaccine development would support them to increase social contact and activities.

Although the initial stages of the COVID-19 pandemic were associated with high levels of psychological distress in general and other disease-specific populations,^{18 19} this study reflects the additional burdens experienced by patients with NMDs and their families. Patients and families demonstrated resilience during the pandemic despite severe limitations on their healthcare-related, academic/professional and social activities, and were able to draw on their prior experiences of adapting to circumstances due to their or their child's condition. In addition, our findings support previous studies reporting that psychological resilience is likely to be greater in those with more perceived care and support, for example, from family and friends.²⁰ It is now apparent that despite the success of the vaccination programme, COVID-19 will be prevalent for some time and healthcare services will need to evaluate both patient preference and health-related outcomes

when planning future services, and it is neither feasible nor fair to ask patients with NMDs, or their families and carers, to shield indefinitely.

This study, our clinical experience, and the limited available body of evidence suggest that COVID-19, particularly with severe outcomes, was uncommon in patients with NMD during and after the first pandemic wave: in a Spanish paediatric NMD registry, 29 individuals tested positive for SARS-CoV-2 to November 2020: 10% were hospitalised and 3% were admitted to ICU.¹³ In addition, in a cohort of 56 patients with spinal muscular atrophy in Hubei province, only one developed COVID-19,²¹ and in a small study of 7 patients with Duchenne muscular dystrophy and COVID-19 infection confirmed on PCR testing, no patients developed moderate–severe disease despite complications including cardiomyopathy and respiratory failure.¹²

Anecdotally we have observed that the rates of COVID-19 increased in our patient cohorts during the second pandemic wave: causes are likely to be multifactorial including the general relaxation of strict shielding measures observed at the start of the pandemic, as well as potentially more transmissible viral mutations. However, very few patients have required hospitalisation and we are unaware of any patient with NMD not being admitted to ICU where this was indicated an expressed preference, although this was a frequently reported concern in this study. In the UK, around 10% of all patients with COVID-19 admitted to ICU care have pre-morbidly required some or total assistance with daily activities²⁰; however, it is possible that those with more severe limitations were deemed clinically inappropriate for escalation or had pre-established ceilings of care. News reports of strain on the NHS is likely to be of greater concern to vulnerable groups as many will have previous experience or have been informed they are more likely to need ICU support in the event of severe illness.

In addition, there was lack of individualised medical advice provided at the start of the pandemic. The lack of clarification from news reports and often medical professionals, for example, that the CFS was not to be used in adults ≤ 65 years with learning or physical disabilities,²² is also likely to have compounded anxieties. There may be a benefit in providing patients and families with individualised action plans to inform emergency care and support, which should also prompt discussion about escalation plans and back-up plans for personal and nursing care.

There was significant change and disruption to the clinical and support services available for patients with NMD during the pandemic.^{23 24} This included an increase in outreach and remote support and consultations, concomitant with a reduction in face-to-face appointments due to reduced service capacity and reluctance of patients to attend hospital due to the risk of nosocomial COVID-19 infection. This posed numerous challenges in the continuation of treatment for NMDs including the reduced availability of adjuncts for ventilatory support, limited provision of supportive therapies, for example, chest

physiotherapy and nutritional management, and delay or cessation of treatments such as corticosteroids.^{23 25}

Remote access to clinical services was viewed positively as an interim measure, but as identified in other groups, this was predominantly for stable patients and problems not considered to require physical examination or diagnostic tests.^{26 27} There remains a paucity of evidence evaluating the clinical outcomes of remote approaches compared with usual standards of care in patients with NMD.²⁸ Perceived discrimination and disadvantage experienced by people with disabilities has been brought into sharp focus during the pandemic in clinical and non-clinical settings: for some patients and families the increased provision of remote services supported their engagement with healthcare services. It is essential for services to consider the aspects highlighted, such as accessibility, in future planning, in which a hybrid model of remote and in person clinical contact may be beneficial.

This study has several limitations. First, the findings reported in this study may not be generalisable beyond the study population. In addition, the study was conducted over several months, during which time the local and national rates of COVID-19 transmission varied as did the availability and content of general and disease-specific guidance. This includes the subsequent availability of vaccinations for vulnerable patients which may have further reduced psychosocial burdens for many. Data collection using telephone questionnaires aimed to maximise the number of people able to take part as it mitigated potential issues with digital exclusion and did not require participants to attend the study site, and provided respondents and researchers the opportunity to clarify and confirm information. However, the study may be limited by selection bias with less representation of those at extremes of the clinical spectrum and those less concerned about COVID-19. In addition, we were unable to administer the questionnaire directly to paediatric patients, and the perspectives of patients are contextually different to those of their parents who responded on their behalf: this means interpretations of the psychosocial impact, including the mental and emotional well-being of their child may differ to that of the patient themselves. Finally, it is important to recognise the potential bias introduced by researchers involved in this study who are all clinicians working with patients from the study population, including some who participated; effects may include respondents avoiding providing negative statements particularly about healthcare services, however a sense of prior familiarity between researchers and patients/parents may have enabled more openness in participants' responses.

CONCLUSIONS

Measures to reduce transmission of COVID-19 have disproportionately affected patients with NMDs and their families, however it is likely that negative psychosocial impacts have and will continue to improve. It is

essential that healthcare professionals continue to evaluate ongoing risk mitigating strategies employed by individuals, and provide up-to-date, accessible and consistent information to advise and support physical and mental well-being. Future research could formally explore the effect on psychosocial outcomes from pre-existing anxiety, depression, socioeconomic status and financial support. In addition, given the dynamic nature of the pandemic, exploring psychosocial impact over different time periods and subsequent pandemic waves, and the impact of vaccination may be of interest.

Contributors LS, H-LT, RW, MC and AS contributed to the planning of the study. LS, H-LT, RW and AS contributed to the conduct of the study. ZH is a patient representative who provided invaluable insight into the significance and interpretation of the results. All authors contributed to the reporting of the study. AS is responsible for the overall content as guarantor. AS takes responsibility for the integrity of the data and the accuracy of the data analysis and the conduct of the study, had full access to all the data and controlled the decision to publish. The corresponding author attests that all listed authors meet authorship criteria and that no others meeting the criteria have been omitted.

Funding This work was supported by the Brompton Breathers Trust Fund, Royal Brompton Hospital; there is no applicable grant number.

Competing interests All authors have completed the Unified Competing Interest form (available on request from the corresponding author) and declare: no support from any organisation for the submitted work; MC reports personal fees from Honorarium paid for lecture at virtual ERS 2020 from ResMed, grants from Support from Breas Medical to attend the ERS 2019, outside the submitted work; and MC also works part-time for Breas Medical. Breas Medical had no part in the development or writing of this manuscript and all time spent on this project was outside of her employment hours for Breas Medical. No other authors have any financial or other relationships or activities that could appear to have influenced the submitted work.

Patient and public involvement Patients and/or the public were involved in the design, or conduct, or reporting, or dissemination plans of this research. Refer to the Methods section for further details.

Patient consent for publication Not applicable.

Ethics approval This study involves human participants and was approved by North West—Greater Manchester Central Research Ethics Committee REC 20/NW/0355. Participants gave informed consent to participate in the study before taking part.

Provenance and peer review Not commissioned; externally peer reviewed.

Data availability statement Data are available upon reasonable request. De-identified participant data may be available upon reasonable request from the corresponding author.

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Author note The lead author (AS) confirms that the manuscript is an honest, accurate and transparent account of the study being reported, that no important aspects of the study have been omitted and that any discrepancies from the study as planned and registered have been explained.

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