Appendix 1. Survey: Jan - Feb 2018

Impact of SMA - all Types 1 - 3

	A	B	C	D	E	F	G	H
1	**Part 1. The impact of SMA - overview - completed once only by either the person or their parent/main unpaid carer**							
2								
3	**1. Completed by:**							
4								
5		**%**	**Nos.**					
6	The unpaid carer (e.g. parent, partner, foster parent) of someone wh	53	69					
7	An adult (age 18 year +) who has SMA	39	51					
8	A young person (age < 18 years) who has SMA	8	10					
9	**Total**	**100**	**130**					
10	Number of returns (one completed jointly)		**128**					
11								
12	**2. Age of the person with SMA:**							
13								
14		**%**	**Nos**					
15	0 - 2 years	8.6	11					
16	3 - 4 years	9.4	12					
17	5 - 12 years	21.9	28					
18	13 - 17 years	11.7	15					
19	18 - 25 years	17.2	22					
20	26 - 35 years	9.4	12					
21	36 - 45 years	6.3	8					
22	46 - 55 years	7.0	9					
23	56 - 65 years	3.9	5					
24	66 + years	4.7	6					
25		**100.0**	**128**					
26								
27								
28								
29								
30								

Appendix 1. Survey: Jan - Feb 2018

Impact of SMA - all Types 1 - 3

	A	B	C	D	E	F	G	H
31	**3. The ethnicity that best describes the person with SMA (as many as apply ticked):**							
32								
33		**%**	**Nos**					
34	English / Scottish / Welsh / Northern Irish / UK	71.43	95					
35	Irish	0.75	1					
36	Any other white background	9.02	12					
37	Indian	3.76	5					
38	Pakistani	4.51	6					
39	Chinese	0.75	1					
40	Any other Asian Background	0.75	1					
41	Arab	0.75	1					
42	Any other ethnic group	0.75	1					
43	Mixed ethnic background	7.52	10					
44		**100.00**	**133**					
45								
46	**4. Type of 5q SMA that affects the person with SMA:**							
47								
48		%	Nos					
49	Type 1	2.3	3					
50	Type 1 / 2	5.5	7					
51	Type 2	53.9	69					
52	Type 2 / 3	7.8	10					
53	Type 3	30.5	39					
54		**100.0**	**128**					
55								
56								
57								
58								
59								
60								

Note: due to the availability of nusinersen via the Type 1 Expanded Access Programme, we did not hear from parents of infants with Type 1 about the impact of the condition when untreated.

Appendix 1. Survey: Jan - Feb 2018

Impact of SMA - all Types 1 - 3

	A	B	C	D	E	F	G	H
61	**5. Health-related impacts that affect the person with SMA:**							
62								
63		**All**	**Type 1-2 age 0 -17**	**Type 1-2 age 18+**	**Type 2/3 - 3 age 0 - 17**	**Type 2/3 - 3 age 18+**		
64	**Numbers**	**128**	**55**	**24**	**11**	**38**		
65		**%**	**%**	**%**	**%**	**%**		
66	Contractures (tightness around joints)	84	87	92	91	71		
67	Pain	62	53	75	73	63		
68	Spinal scoliosis (curvature)	60	65	75	36	50		
69	Prolonged mealtimes / fatigue with oral feeding	50	56	71	18	37		
70	Constipation	45	60	25	27	42		
71	Osteopenia (bone weakness due to bone density)	41	29	67	27	45		
72	Shortness of breath	40	38	42	36	42		
73	Chest infections / pneumonia	34	35	42	18	32		
74	Excessive secretions	31	31	12	18	24		
75	Choking issues or coughing during swallowing	30	22	58	18	29		
76	Nausea / morning headaches / frequent night time waking (due to lo	25	22	29	27	26		
77	Insufficient weight gain	20	33	6	9	3		
78	Excessive weight gain	19	11	3	18	34		
79	Bone fractures	14	5	5	18	21		
80	Gastro-oesophageal reflux	13	11	6	0	11		
81								
82								
83								
84								
85								
86								
87								
88								
89								

Appendix 1. Survey: Jan - Feb 2018

Impact of SMA - all Types 1 - 3

	A	B	C	D	E	F	G	H
90	**6. Health-related interventions and equipment currently used by the person with SMA - both NHS and non-NHS funded**							
91								
92		**All**	**Type 1-2 age 0 -17**	**Type 1-2 age 18+**	**Type 2/3 - 3 age 0 - 17**	**Type 2/3 - 3 age 18+**		
93	**Numbers**	**128**	**55**	**24**	**11**	**38**		
94		**%**	**%**	**%**	**%**	**%**		
95	Powered wheelchair	83	93	92	36	76		
96	Adaptations to toilet and bathroom facilities	73	62	83	55	92		
97	Other home adaptations (e.g. bathroom, kitchen, ramps, lift)	69	60	83	27	84		
98	Manual wheelchair	68	76	71	64	55		
99	Wheelchair accessible vehicle	66	64	83	45	63		
100	Specialist bed	63	62	83	45	55		
101	Hoist (mobile or ceiling track)	60	60	90	36	47		
102	Orthotics (e.g. leg splints)	54	85	38	73	13		
103	Specialist seating	50	64	63	55	21		
104	Medication e.g. for constipation	48	65	42	36	29		
105	Salbutamol	46	53	50	73	26		
106	Chest physiotherapy	43	64	46	27	16		
107	Assisted cough (e.g. cough assist machine)	38	53	92	0	16		
108	Spinal rods / spinal fusion	35	22	54	9	29		
109	Standing frame	33	65	0	27	8		
110	Nebuliser	31	42	58	0	11		
111	Assistive technology	30	31	46	18	24		
112	Night time non-invasive ventilation	29	23	58	0	8		
113	Oral suctioning	26	40	38	0	5		
114	Spine orthotics (spinal brace / jacket)	20	19	25	9	0		
115	Specialist car seat	18	31	4	18	8		
116	Wizzybug	16	33	0	27	0		
117	Gastrostomy	16	24	29	9	0		

Appendix 1. Survey: Jan - Feb 2018
Impact of SMA - all Types 1 - 3

	A	B	C	D	E	F	G	H
118	Walking frame	15	18	0	45	11		
119	Postural draining	14	15	25	0	11		
120	Mobile arm supports	9	18	4	0	3		
121	Day time non-invasive ventilation	9	9	25	0	3		
122	Lycra suit	7	15	4	0	0		
123	Mechanical ventilation / tracheostomy	2	5	0	0	0		
124	Tube feeding (ng or nj)	2	4	4	0	0		
125								
126	**7. Health-related interventions and equipment used by the person with SMA - not NHS-funded:**							
127								
128		**As % of those who use this**	**As a % of all respondents**					
129	**Equipment / intervention**		**All**	**Type 1-2 age 0 -17**	**Type 1-2 age 18+**	**Type 2/3 - 3 age 0 - 17**	**Type 2/3 - 3 age 18+**	
130		**%**	**%**	**%**	**%**	**%**	**%**	
131	Wizzybug	71	15	31	0	18	0	
132	Specialist car seat	70	16	27	4	18	8	
133	Tube feeding (ng or nj)	67	1	2	4	0	0	
134	Wheelchair accessible vehicle	57	48	47	67	18	47	
135	Other home adaptations (e.g. bathroom, kitchen, ramps, lift)	52	46	33	63	0	68	
136	Powered wheelchair	50	41	53	46	0	34	
137	Assistive technology	50	19	13	29	9	24	
138	Postural draining	44	8	9	21	0	0	
139	Mobile arm supports	42	5	11	0	0	3	
140	Adaptations to toilet and bathroom facilities	36	47	31	63	0	71	
141	Lycra suit	33	2	5	4	0	0	
142	Mechanical ventilation / tracheostomy	33	1	2	0	0	0	
143	Hoist (mobile or ceiling track)	27	21	18	54	0	11	
144	Walking frame	26	2	5	0	0	8	
145	Specialist bed	24	19	9	33	9	26	
146	Chest physiotherapy	24	12	18	25	0	0	

Appendix 1. Survey: Jan - Feb 2018

Impact of SMA - all Types 1 - 3

	A	B	C	D	E	F	G	H
147	Manual wheelchair	23	20	22	13	18	24	
148	Specialist seating	20	13	15	17	0	13	
149	Nebuliser	20	8	11	17	0	0	
150	Day time non-invasive ventilation	17	1	2	4	0	0	
151	Gastrostomy	10	2	2	4	0	0	
152	Medication e.g. for constipation	8	5	5	0	0	8	
153	Spine orthotics (spinal brace / jacket)	8	2	2	4	0	0	
154	Assisted cough (e.g. cough assist machine)	6	3	5	4	0	0	
155	Oral suctioning	6	2	4	0	0	0	
156	Standing frame	5	3	4	0	0	3	
157	Spinal rods / spinal fusion	4	2	2	4	0	3	
158	Night time non-invasive ventilation	3	1	2	0	0	0	
159	Salbutamol	2	1	0	0	0	3	
160	Orthotics (e.g. leg splints)	1	1	0	4	0	0	
161	**Numbers of respondents**		**128**	**55**	**24**	**11**	**38**	
162								
163	**8. Health-related interventions and equipment the person with SMA does not have but wants:**							
164								
165		**All**	**Type 1-2 age 0 -17**	**Type 1-2 age 18+**	**Type 2/3 - 3 age 0 - 17**	**Type 2/3 - 3 age 18+**		
166	**Numbers**	**128**	**55**	**24**	**11**	**38**		
167		**%**	**%**	**%**	**%**	**%**		
168	Other home adaptations (e.g. bathroom, kitchen, ramps, lift)	15	24	4	36	3		
169	Adaptations to toilet and bathroom facilities	14	24	0	45	0		
170	Chest physiotherapy	13	15	21	0	8		
171	Assistive technology	13	22	17	0	3		
172	Wheelchair accessible vehicle	12	20	4	9	5		
173	Mobile arm supports	10	18	0	0	0		
174	Specialist bed	9	18	4	0	0		

Appendix 1. Survey: Jan - Feb 2018

Impact of SMA - all Types 1 - 3

	A	B	C	D	E	F	G	H
175	Hoist (mobile or ceiling track)	9	16	0	9	3		
176	Powered wheelchair	8	7	4	9	8		
177	Specialist car seat	8	16	0	9	0		
178	Walking frame	8	16	0	9	0		
179	Lycra suit	8	16	0	0	3		
180	Assisted cough (e.g. cough assist machine)	7	9	13	0	3		
181	Specialist seating	6	9	8	9	0		
182	Spine orthotics (spinal brace / jacket)	6	13	0	0	3		
183	Manual wheelchair	5	7	4	18	0		
184	Standing frame	5	5	0	0	3		
185	Nebuliser	5	9	0	0	3		
186	Salbutamol	4	4	13	0	0		
187	Day time non-invasive ventilation	4	7	0	0	0		
188	Night time non-invasive ventilation	3	5	4	0	0		
189	Wizzybug	3	7	0	0	0		
190	Orthotics (e.g. leg splints)	2	2	8	0	0		
191	Medication e.g. for constipation	2	4	0	0	0		
192	Spinal rods / spinal fusion	2	4	4	0	0		
193	Oral suctioning	2	4	4	0	0		
194								
195								
196								
197								
198								
199								
200								
201								
202								
203								
204								

Appendix 1. Survey: Jan - Feb 2018

Impact of SMA - all Types 1 - 3

	A	B	C	D	E	F	G	H
205								
206	**9. The number of health & social care professionals seen by the person with SMA / carer in connection with the SMA in the last 12 months**							
207								
208		**All**	**Type 1-2 age 0 -17**	**Type 1-2 age 18+**	**Type 2/3 - 3 age 0 - 17**	**Type 2/3 - 3 age 18+**		
209	**Numbers**	**128**	**55**	**24**	**11**	**38**		
210		**%**	**%**	**%**	**%**	**%**		
211	0 - 5 professionals	37	15	29	18	79		
212	6 - 10 professionals	32	33	46	55	16		
213	11 - 20 professionals	25	45	21	18	0		
214	21+ professionals	6	7	4	9	5		
215		100	100	100	100	100		
216								
217	**10. Daily living tasks for which the person with SMA needs full support:**							
218								
219		**All**	**Type 1-2 age 0 -17**	**Type 1-2 age 18+**	**Type 2/3 - 3 age 0 - 17**	**Type 2/3 - 3 age 18+**		
220	**Numbers**	**128**	**55**	**24**	**11**	**38**		
221		**%**	**%**	**%**	**%**	**%**		
222	Dressing	81	96	96	64	55		
223	Transfers	80	100	96	64	47		
224	Toileting	78	100	96	64	39		
225	Preparing meals	75	93	83	64	47		
226	Washing	74	91	92	55	45		
227	Eating and drinking	31	38	58	9	11		
228								
229								
230								

Appendix 1. Survey: Jan - Feb 2018

Impact of SMA - all Types 1 - 3

	A	B	C	D	E	F	G	H
231								
232	**11. Number of people with SMA needing night care (e.g. help to turn over at night):**							
233								
234		**All**	**Type 1-2 age 0 -17**	**Type 1-2 age 18+**	**Type 2/3 - 3 age 0 - 17**	**Type 2/3 - 3 age 18+**		
235	**Numbers**	**128**	**55**	**24**	**11**	**38**		
236		**%**	**%**	**%**	**%**	**%**		
237	Yes	66	85	92	73	47		
238								
239	**12. Average number of times a night those needing night care need it**							
240								
241		**All**	**Type 1-2 age 0 -17**	**Type 1-2 age 18+**	**Type 2/3 - 3 age 0 - 17**	**Type 2/3 - 3 age 18+**		
242	**Numbers**	**99**	**52**	**22**	**7**	**18**		
243		**%**	**%**	**%**	**%**	**%**		
244	1 - 2 times	36	21	37	86	55.5		
245	3 - 4 times	26	21	37	14	33		
246	5 - 6 times	22	33	18	0	5.5		
247	6+ times	16	25	9	0	5.5		
248		100	100	100	100	100		
249								
250								
251								
252								
253								
254								
255								
256								

Appendix 1. Survey: Jan - Feb 2018

Impact of SMA - all Types 1 - 3

	A	B	C	D	E	F	G	H
257								
258	**13. Average hours of H and SS paid personal assistance the person/unpaid carer has for each day / night (24 hour) period:**							
259								
260		**All**	**Type 1-2 age 0 -17**	**Type 1-2 age 18+**	**Type 2/3 - 3 age 0 - 17**	**Type 2/3 - 3 age 18+**		
261	**Numbers**	**128**	**55**	**24**	**11**	**38**		
262		**%**	**%**	**%**	**%**	**%**		
263	0 hours	48	58	4	64	55		
264	1 - 5 hours	19	17	12	9	13		
265	6 - 10 hours	6	11	0	9	16		
266	11 - 20 hours	7	7	16	9	3		
267	21 - 24 hours	20	7	68	9	13		
268		100	100	100	100	100		
269								
270	**14. Who provides any unpaid assistance needed by the person with SMA:**							
271								
272		**All**	**Type 1-2 age 0 -17**	**Type 1-2 age 18+**	**Type 2/3 - 3 age 0 - 17**	**Type 2/3 - 3 age 18+**		
273	**Numbers**	**128**	**55**	**24**	**11**	**38**		
274		**%**	**%**	**%**	**%**	**%**		
275	Parent	75	91	79	9	47		
276	Other relatives	33	51	13	4	18		
277	Partner	12	0	8	0	34		
278	Friends	17	9	29	1	24		
279	I don't have any	10	0	21	0	21		
280	Son / daughter	5	0	4	0	16		
281								
282								
283								

Appendix 1. Survey: Jan - Feb 2018

Impact of SMA - all Types 1 - 3

	A	B	C	D	E	F	G	H
284								
285	**15. The other caring responsibilities the main unpaid carer / assistant has:**							
286								
287		**All**	**Type 1-2 age 0 -17**	**Type 1-2 age 18+**	**Type 2/3 - 3 age 0 - 17**	**Type 2/3 - 3 age 18+**		
288	**Numbers**	**128**	**55**	**24**	**11**	**38**		
289		**%**	**%**	**%**	**%**	**%**		
290	Children	51	64	39	55	37		
291	Ageing parents	32	29	50	36	27		
292	None	22	11	33	18	27		
293	Other relatives	20	33	11	9	13		
294								
295	**Q16. How many of the unpaid carers / assistants have given up work to assist the person with SMA:**							
296								
297		**All**	**Type 1-2 age 0 -17**	**Type 1-2 age 18+**	**Type 2/3 - 3 age 0 - 17**	**Type 2/3 - 3 age 18+**		
298	**Numbers responding**	**114**	**54**	**20**	**11**	**30**		
299	**Number of unpaid carers**	**146**	**76**	**25**	**12**	**37**		
300		**%**	**%**	**%**	**%**	**%**		
301	Yes - completely	39	45	36	42	27		
302	Yes - had to drop to part time	25	29	16	50	19		
303								
304								
305								
306								

Appendix 1. Survey: Jan - Feb 2018

Impact of SMA - all Types 1 - 3

	A	B	C	D	E	F	G	H
307	**Part 2. The impact of SMA - views of the person, their parent/main unpaid carer and relatives**							
308								
309	**1. Are the interventions and support the person with SMA and their unpaid carer(s) have in their day-to-day lives enough for the person with SMA to manage and..**							
310								
311		**All**	**Type 1-2 age 0 -17**	**Type 1-2 age 18+**	**Type 2/3 - 3 age 0 - 17**	**Type 2/3 - 3 age 18+**		
312	**Numbers of people with SMA giving their view**	56	12	14	3	27		
313	**Numbers of unpaid carers giving their view**	55	28	7	7	13		
314	**Numbers of relatives giving their view**	21						
315	**Total views**	**132**	**40**	**21**	**10**	**40**		
316		**No/Not really**						
317		%	%	%	%	%		
318	Get out and about	60	50	33	70	70		
319	Keep physically well	58	43	52	60	70		
320	Keep emotionally well	56	43	62	70	58		
321	Work / study for the hours you wish	52	43	76	50	50		
322	Keep socially connected	54	53	29	30	68		
323	Get enough sleep	54	55	62	50	43		
324	Manage financially	45	40	43	30	48		
325								

Appendix 1. Survey: Jan - Feb 2018

Impact of SMA - all Types 1 - 3

	A	B	C	D	E	F	G	H
326	**2. Are the interventions and support the person with SMA and their unpaid carer(s) have in their day-to-day lives enough for the main unpaid carer / unpaid personal assistant to manage and..**							
327								
328		**All**	**Type 1-2 age 0 -17**	**Type 1-2 age 18+**	**Type 2/3 - 3 age 0 - 17**	**Type 2/3 - 3 age 18+**		
329	**Numbers of people with SMA giving their view**	56	12	14	3	27		
330	**Numbers of unpaid carers giving their view**	55	28	7	7	13		
331	**Numbers of relatives giving their view**	21						
332	**Total views**	**132**	**40**	**21**	**10**	**40**		
333		**No/not really**						
334		**%**	**%**	**%**	**%**	**%**		
335	Get enough sleep	73	83	62	70	58		
336	Work /study for the hours they wish	70	78	62	90	50		
337	Keep physically well	67	70	52	40	63		
338	Keep emotionally well	67	73	57	70	53		
339	Manage financially	60	65	38	70	58		
340	Get out and about	57	60	38	50	60		
341	Keep socially connected	54	50	43	60	58		