

SMA Adult Outcomes Delphi Process Round 1 Report

The first round of the Delphi Process for SMA has been completed. Below contains a brief report including figures and tables summarizing the results of the first round of the Delphi Process, as a breakdown of outcomes. The major assessments for examining consensus were the averages, and confidence intervals for each outcome. Tables 1 and 2 describe outcomes which have been favourably rated, by examining the confidence intervals or averages.

Table 3 includes all averages, standard deviations and confidence intervals.

In total, there were 10 responses to the Round 1 survey, figure 1 outlines the provinces and specialties of each of the respondents. Outcomes included in round 2 were outcomes which met at least a rank of 3 (Neither strong nor weak) in the previous round. This narrowed the list of outcomes from 34 to 21.

When completing the second-round survey, please utilize the attached outcome table if needed. This outcome table includes valuable information regarding each of the outcomes being considered.

When ranking outcomes, we are looking for your opinions on which outcomes are the best outcomes; considering the quality, and utility of the outcomes and not necessarily the outcomes that are currently being used. The second round of voting will be open until approximately October 25th, 2019.

Once voting is complete, we will again compile the results and anonymously release the results to the group. Depending on whether a consensus was reached or not, a third round of voting could begin.

Updates to Round 2:

1. There is a reduced pool of outcomes following the first round, there is now only 21 outcomes included in Round 2, down from a total of 34. The included outcomes are outcomes that met a cut off of an average score of 3+. Additional cut points are available in the report of Round 1, which is available via email.
2. There is now an additional option to rate an outcome as unknown - this rating will be exempt from the analysis and will provide higher quality data.
3. There are additional open-ended questions, looking to explore why rankings are made the way they are, whether you are currently using the outcomes, and if there are any additional comments for the outcomes to include. This additional information will help guide the next round of results.

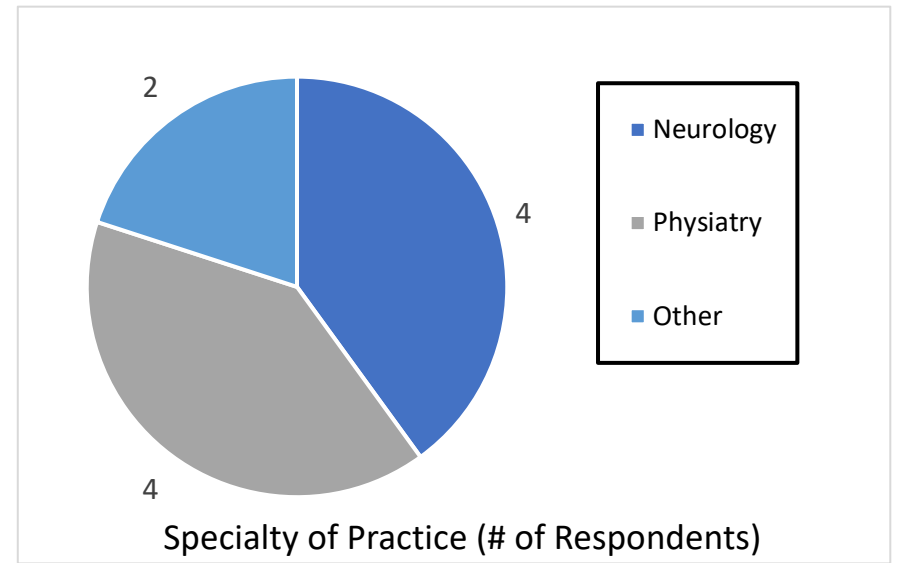
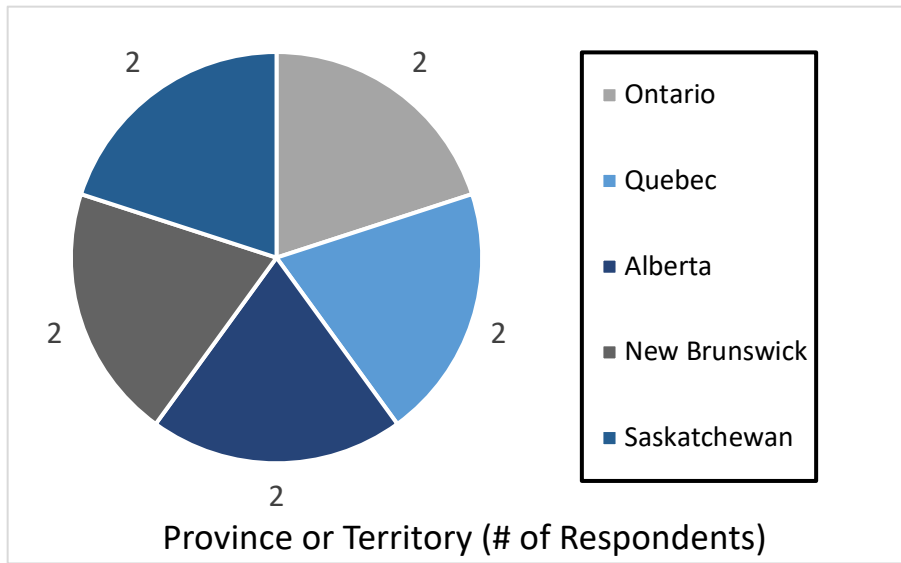


Figure 1. Geographic Breakdown (Left) and Speciality of Practice (Right) of Respondents

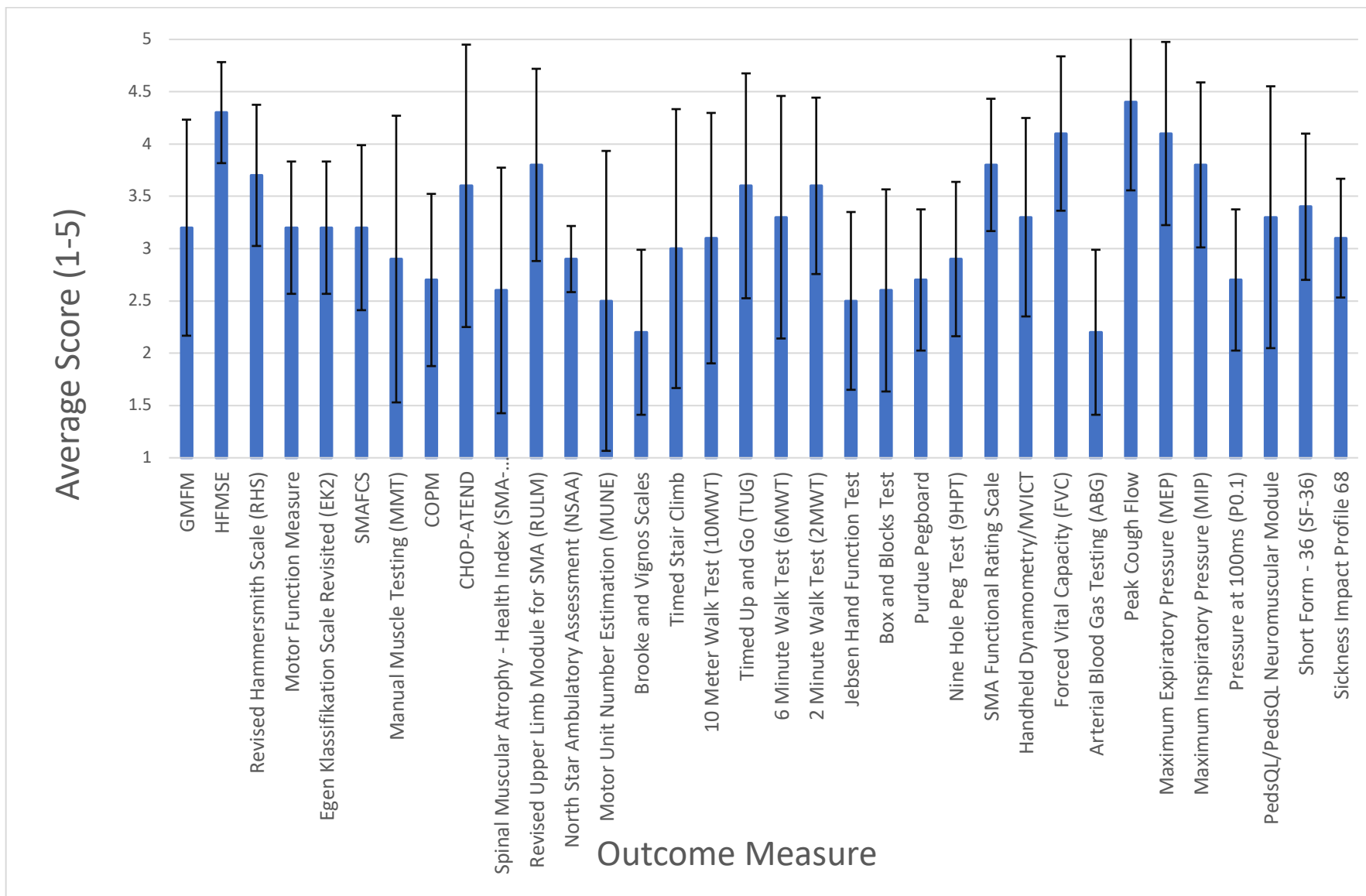


Figure 2. Average Scores of outcome measures (error bars indicate Standard Deviation)

Table 1. Outcomes which achieved Confidence intervals with lower bounds above 3 (Neither Weak nor Strong) or 3.5

Confidence Intervals	
Lower Bound Greater than 3 (N = 9)	Lower Bound Greater than 3.5 (N = 4)
Hammersmith Functional Motor Scale - Expanded	Hammersmith Functional Motor Scale - Expanded
Revised Hammersmith Scale (RHS)	Forced Vital Capacity (FVC)
Revised Upper Limb Module for SMA (RULM)	Peak Cough Flow
2 Minute Walk Test (2MWT)	Maximum Expiratory Pressure (MEP)
SMA Functional Rating Scale	
Forced Vital Capacity (FVC)	
Peak Cough Flow	
Maximum Expiratory Pressure (MEP)	
Maximum Inspiratory Pressure (MIP)	

Table 2. Outcomes which achieved averages of greater than 3, 3.5, and 4 respectively.

Average		
Greater than 3 (N=21)	Greater than 3.5 (N=11)	Greater than 4 (N=4)
Gross Motor Function Measure (GMFM)	Hammersmith Functional Motor Scale - Expanded	Hammersmith Functional Motor Scale - Expanded
Hammersmith Functional Motor Scale - Expanded	Revised Hammersmith Scale (RHS)	Forced Vital Capacity (FVC)
Revised Hammersmith Scale (RHS)	CHOP-ATEND	Peak Cough Flow
Motor Function Measure	Revised Upper Limb Module for SMA (RULM)	Maximum Expiratory Pressure (MEP)
Egen Klassifikation Scale Revisited (EK2)	Timed Up and Go (TUG)	
Spinal Muscular Atrophy Functional Composite Scale (SMAFCS)	2 Minute Walk Test (2MWT)	
CHOP-ATEND	SMA Functional Rating Scale	
Revised Upper Limb Module for SMA (RULM)	Forced Vital Capacity (FVC)	
10 Meter Walk Test (10MWT)	Peak Cough Flow	
Timed Up and Go (TUG)	Maximum Expiratory Pressure (MEP)	
6 Minute Walk Test (6MWT)	Maximum Inspiratory Pressure (MIP)	
2 Minute Walk Test (2MWT)		
SMA Functional Rating Scale		
Handheld Dynamometry/MVICT		
Forced Vital Capacity (FVC)		
Peak Cough Flow		
Maximum Expiratory Pressure (MEP)		
Maximum Inspiratory Pressure (MIP)		
PedsQL/PedsQL Neuromuscular Module		
Short Form - 36 (SF-36)		
Sickness Impact Profile 68		

Table 3. Averages and Confidence Intervals of each of the assessed outcome measures

	Average [Standard Deviation]	Confidence Interval
Gross Motor Function Measure (GMFM)	3.2 [1.03]	2.54-3.85
Hammersmith Functional Motor Scale - Expanded	4.3 [0.48]	3.99-4.60
Revised Hammersmith Scale (RHS)	3.7 [0.67]	3.27-4.12
Motor Function Measure	3.2 [0.63]	2.8-3.6
Egen Klassifikation Scale Revisited (EK2)	3.2 [0.63]	2.8-3.6
Spinal Muscular Atrophy Functional Composite Scale (SMAFCS)	3.2 [0.78]	2.70-3.69
Manual Muscle Testing (MMT)	2.9 [1.37]	2.03-3.76
Canadian Occupational Performance Measure (COPM)	2.7 [0.82]	2.17-3.22
CHOP-ATEND	3.6 [1.34]	2.74-4.45
Spinal Muscular Atrophy - Health Index (SMA-HI)	2.6 [1.17]	1.85-3.34
Revised Upper Limb Module for SMA (RULM)	3.8 [0.91]	3.21-4.38
North Star Ambulatory Assessment (NSAA)	2.9 [0.31]	2.7-3.1
Motor Unit Number Estimation (MUNE)	2.5 [1.43]	1.59-3.40
Brooke and Vignos Scales	2.2 [0.78]	1.70-2.69
Timed Stair Climb	3 [1.33]	2.15-3.84
10 Meter Walk Test (10MWT)	3.1 [1.19]	2.34-3.85
Timed Up and Go (TUG)	3.6 [1.07]	2.92-4.27
6 Minute Walk Test (6MWT)	3.3 [1.15]	2.56-4.03
2 Minute Walk Test (2MWT)	3.6 [0.84]	3.06-4.13
Jebsen Hand Function Test	2.5 [0.84]	1.96-3.03
Box and Blocks Test	2.6 [0.96]	1.98-3.21
Purdue Pegboard	2.7 [0.67]	2.27-3.12
Nine Hole Peg Test (9HPT)	2.9 [0.73]	2.43-3.36
SMA Functional Rating Scale	3.8 [0.63]	3.4-4.2
Handheld Dynamometry/Maximum Voluntary Isolation Contraction Testing	3.3 [0.94]	2.7-3.9
Forced Vital Capacity (FVC)	4.1 [0.73]	3.63-4.56
Arterial Blood Gas Testing (ABG)	2.2 [0.78]	1.70-2.69
Peak Cough Flow	4.4 [0.84]	3.86-4.93
Maximum Expiratory Pressure (MEP)	4.1 [0.87]	3.54-4.65
Maximum Inspiratory Pressure (MIP)	3.8 [0.78]	3.30-4.29
Pressure at 100ms (P0.1)	2.7 [0.67]	2.27-3.12
PedsQL/PedsQL Neuromuscular Module	3.3 [1.25]	2.50-4.09
Short Form - 36 (SF-36)	3.4 [0.69]	2.95-3.84
Sickness Impact Profile 68	3.1 [0.56]	2.74-3.45