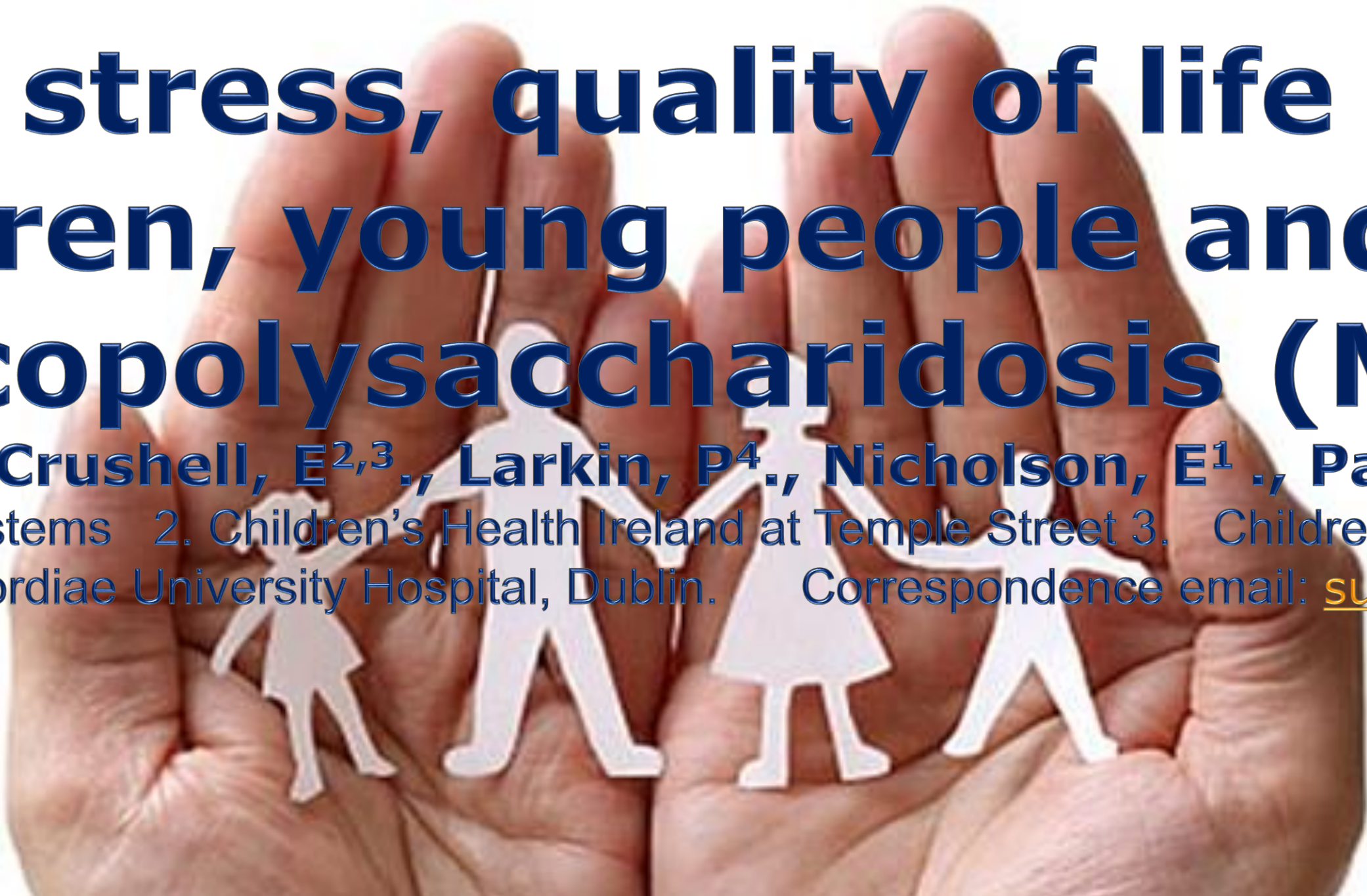


# Measuring parenting stress, quality of life and resilience related to caring for children, young people and adults living with Mucopolysaccharidosis (MPS)

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## Background

- Mucopolysaccharidosis (MPS) are a rare group of inherited metabolic disorders.
- A defective or missing specific lysosomal enzyme causes large amounts of complex sugar molecules or glycoaminoglycins (GAGS) to accumulate in harmful amounts in the body's cells and tissues.
- This results in progressive cellular damage, which in turn leads to an array of manifestations that worsen with age, and can affect multiple organ systems, leading to cognitive impairment and eventually resulting in severe morbidity and premature death (Muenzer, 2014; Giugliani, 2012; Muenzer, 2011; Clarke, 2008; Kircher *et al.* 2007).
- Disease trajectories are often uncertain in children with MPS due to life-threatening or chronically debilitating nature of the conditions, and most of the children required to meet constant caregiving needs.

## Aim and Objectives

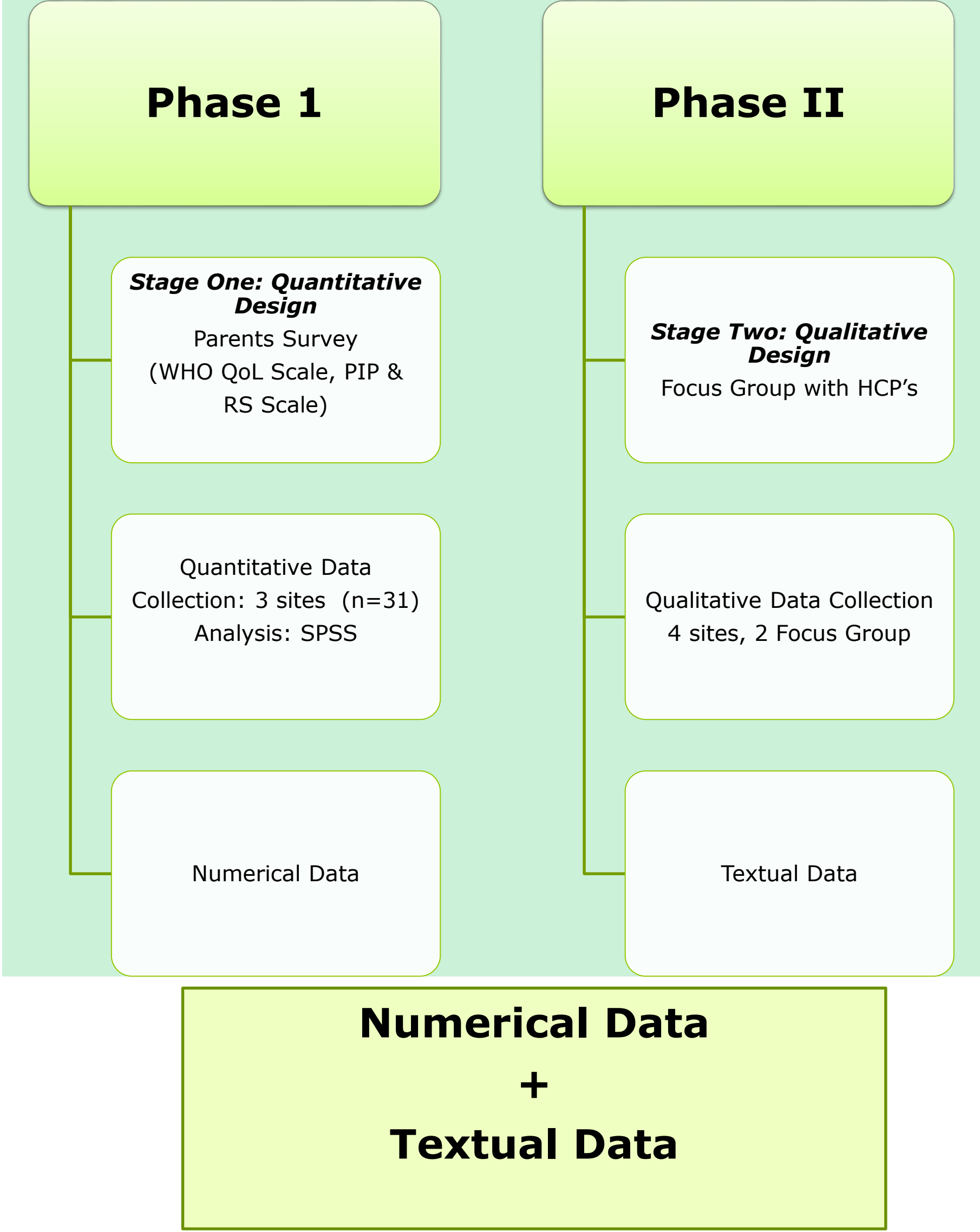
To investigate the impact of caring for children, young people and adults living with MPS from parents’ or caregiver's perspective.

## Ethics

The study has been approved by the Research and Ethics Committee from the three clinical sites and Ethical exemption was sanctioned by the University College Dublin Ethics Board.

## Method

A sequential explanatory design was used in this study.



- Phase 1 cross-sectional study results are presented in this poster.
- A total of 31 participants, all parents or caregivers for patients with MPS (25.8% male and 72.4% females) completed standardised measures of quality of life, parenting stress as it related to children with a long-term illness, and resilience.

## WHOQOL-BREF Scale (WHO, 1996)

	MPS I			MPS II			MPS IV			
Domains	Mean Score	1-100 Transformed Score	% of Max	Mean Score	1-100 Transformed Score	% of Max	Mean Score	1-100 Transformed Score	% of Max	Maximum Possible
Physical health	25.67	69	73.34%	24.2	63	69.14%	29.61	81	84.6%	35
Psychological	23.91	75	79.7%	21.33	63	71.1%	19.8	56	66%	30
Social relationships	12.57	81	83.8%	10.33	56	68.86%	9.8	56	65.33%	15
Environment	31.04	75	77.6%	28.33	63	70.83%	29.6	69	74%	40
Self-reported QoL (Q1)	4.43	-----	88.6%	3.67	-----	73.4%	3.8	-----	76%	5

## Key findings: Quality of Life (QoL)

- Regarding specific domains, the highest score was for physical health (Domain 1) with a pooled mean of 76.19, compared to the lowest score, for Domain 2 (psychological health) of 70.23.
- Parents of children with MPS I scored the highest in all domains except for Domain 1. They scored an average of 11.01% above the other cohorts across the four domains.
- MPS IV scored the highest in Domain 1, scoring on average 13.36% above the other typologies. The Cronbach’s alpha of this survey ranged from 0.704 to 0.868.
- The average WHOQOL-BREF score was 46.59 in (Guarany *et al.* 2015) study with similar population in Brazil, with the physical health domain scoring highest and the environmental domain scoring lowest.

## Pediatric Inventory for Parents (PIP)

(Streisand, 2016)

	MPS I		MPS II		MPS IV		
	Mean score	% of Max	Mean Score	% of Max	Mean Score	% of Max	Maximum Possible
CMF	19.45	43.22%	29	64.44%	20.2	44.88%	45
CMD	18.52	41.15%	33.67	74.82%	19	42.22%	45
MCF	19.95	49.88%	30.66	76.65%	22.8	57%	40
MCD	17.73	44.33%	29	72.5%	18.6	46.5%	40
EDF	39.73	48.97%	60	85.7%	40.6	54.13%	75
EDD	40.74	54.32%	66.67	88.89%	41	54.66%	75
RFF	20.38	40.76%	30.3	60.6%	21	42%	50
RFD	22.70	45.4%	39.33	78.66%	22	44%	50
F Total	100.45	47.8%	151	71.9%	104.4	49.71%	210
D Total	97.26	46.3%	167.33	79.7%	102.8	48.9%	210

## Key findings: PIP

- Many of the results show high levels of concern for the future with just under 50% finding themselves very often feeling scared that their child’s condition will deteriorate or that their child will die and report finding these thoughts very difficult.
- 55% of caregivers find themselves worrying about the future of their child’s condition regularly and report feeling helpless regarding the condition.
- MPS II scored the highest in all categories by an average of 25.51%, specifically frequency was 18.88% higher than the other cohorts and difficulty scored 41.92% higher than MPS I and IV. The Cronbach’s alpha of this survey ranged from 0.364 (Communication Frequency) to 0.941 (Emotional Distance Difficulty).

## Resilience Scale

(Wagnild &Young, 1993)

	MPS I		MPS II		MPS IV		
	Mean score	% of Max	Mean Score	% of Max	Mean Score	% of Max	Maximum Possible
Total	81.35	83%	84.33	86%	80	81.6%	98

## Key findings: Resilience Scale

- 32.2% of the sample scored between 64 and 81, who are rated as having high resilience tendencies.
- Only (16.1%) were shown to have average resilience tendencies, scoring between 49 and 63.
- These higher than average scores for resilience can be considered in conjunction with question 26 of the WHOQOL-BREF, where 74.2% of the sample stated they felt negative feelings such as anxiety or depression seldom or never.

## Overall Findings

- ✓ Families with MPS seem to overwhelmingly voice concern about their child’s future.
- ✓ Five of the thirty-one participants reported poor physical health at the time of survey. However, it seems to have very little impact on their overall health, with psychological stressors being the more prevalent issue.
- ✓ The overall mean QoL score from the WQB was 93.81 indicated better quality of life.