

Long-term neuropsychological sequelae, emotional wellbeing and quality of life in patients with acquired thrombotic thrombocytopenic purpura

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SUPPLEMENTARY DATA

Table S1. Enrolment criteria.

	<i>Patient characteristics</i>
<i>Inclusion criteria</i>	(a) diagnosis of acquired TTP*
	(b) 18 years of age or older at the time of enrolment
	(c) adequate Italian language for the purpose of a valid psychometric testing
<i>Exclusion criteria</i>	(a) presence of severe neurological or psychiatric diseases before the onset of TTP
	(b) drug or alcohol-abuse possibly impacting on quality of life and neuropsychological tests

* Acquired TTP was defined by at least one episode of thrombocytopenia and microangiopathic hemolytic anemia, with exclusion of alternative explanations,¹ and by evidence of severe ADAMTS13 deficiency (i.e., < 10% activity) in at least one plasma sample collected during the acute episode or disease remission, and/or by normalization of ADAMTS13 activity levels in the remission phase. ADAMTS13 activity was measured using a modified FRETTS-VWF73 or CBA assay.²

Table S2. Psychological measures.

Assessment	Goal	Test name	Administration	Scoring
Cognitive measure	Short memory, direct memory	Digit span ³	List of numbers that one has to repeat in correct order immediately after presentation.	Cognitive tests are standardized measures with population-based normative data that allow the examiner to evaluate a patient's performance with an appropriate group of reference (e.g., those of the same age group). Normative data are generally gathered on typically healthy subjects who are free from diagnosed cognitive dysfunctions, mental illness, disability or neurological disorders that could affect cognitive performance (all these variables cannot be controlled using a generic control group). Data are typically gathered on samples that reflect the broad demographic characteristics of the country of reference including factors such as age, and educational status. ⁶ Each subject's raw score on each test was converted to a standardized score based on normative data generated from the value of the normal population according to the subject's age and education level, as appropriate. Standardized scores of TTP patients were then compared with norm-referenced data from the Italian population. ⁷⁻⁹
	Short memory; backward memory	Digit span backward ³	List of numbers that one has to repeat in reverse order immediately after presentation.	
	Immediate memory; deferred memory	Word Rey List (direct and indirect) ⁴	Word-list memory task in which 15 unrelated words are presented orally over three consecutive learning trials: the participant is asked to recall as many words as possible, after each presentation (direct score) and after 15 minutes (recall score).	
	Sustained attention, focused attention	Trail Making Test A ⁵	Patient is instructed to connect a set of 25 dots of numbers as fast as possible while still maintaining accuracy.	
	Sustained attention, divided attention	Trail Making Test B ⁵	Patient is instructed to connect a set of numbers and letters as fast as possible while still maintaining accuracy.	
Emotional wellbeing	Anxiety	Hamilton Anxiety Rating Scale ¹⁰	Psychologist-driven interview consisting of 14 items, each defined by a series of symptoms, and measures both psychic anxiety (mental agitation and psychological distress) and somatic anxiety (physical complaints related to anxiety).	Each item is scored on a scale of 0 (not present) to 4 (severe), where ≤ 13 indicates absence of the disorder, 14-17 mild severity, 18-24 moderate severity and ≥ 25 severe anxiety. ¹⁰
	Depression	Hamilton Depression Rating Scale ¹¹	Psychologist-driven interview consisting of 17 items, each defined by a series of symptoms, and measures associated with depression (e.g. insomnia, hyperphagia).	The score varies from ≤ 7 (absence of depression), 8-17 (mild depression), 18-24 (medium depression) and ≥ 25 (severe depression). ¹¹
Health-related quality of life (HrQoL)	HrQoL	SF-36 ^{12,13}	Self-report scale evaluating 8 dimensions: physical activity (PA), role physically (RP), bodily pain (BP), general health (GH), vitality (VI), social functioning (SF), role emotional (ER) and mental health (MH).	Each scale is directly transformed into a 0-100 scale on the assumption that each question carries equal weight (min-max: 0-100). To aid interpretation, norm-based scoring has been introduced for the SF-36, setting the general mean at 50 and the standard deviation (SD) at 10 for all scales, with higher scores indicating better health (i.e., a score of zero is equivalent to maximum disability and a score of 100 is equivalent to the best health status perceived). ¹⁴ Each subject's raw score on each test was converted to a standardized score based on normative data generated from the value of the normal population according to the subject's age and education level, as appropriate. ^{14, 15} Standardized scores of TTP patients were compared with norm referenced data from the Italian population. ¹⁵

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