

Young Onset Dementia and role of Cognitive Reserve

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INTRODUCTION

The aim of the present work is to verify whether Cognitive Reserve (CR) maintains its protective role even in patients with an early diagnosis of dementia. This concept has been extensively investigated in the typical onset of neurodegenerative disease, but it is still poorly considered in patients with young onset (Young Onset Dementia, YOD) which is an important clinical condition because of both medical and social consequences (Sampson, Warren & Rossor, 2004). The diagnosis of a neurodegenerative disease may be difficult when considering protective factors such as CR and the young age of the patient at the onset. These factors may mislead the identification of symptoms, even in the case of strong diagnostic suspicions.

METHODS

Our preliminary study refers to four patients with YOD with an average age of 60.75 years, two of them with very high CR and two with low CR. The level of CR was measured with the Cognitive Reserve Index questionnaire (CRIq, Nucci et al., 2012), a simple and quick instrument that allows to collect information on education, working activities and leisure time activities. Moreover, participants were also assessed through a flexible battery of neuropsychological tests (see figures below for some examples).



PATIENT 1 (FIRST ASSESSMENT)

CM	VALUTAZIONE ATTUALE: 21/01/2016				
TEST	PUNT. GREZZO	CUT-OFF	ESITO		
MMSSE	MMSSE totale grezzo	9/30	24	ALTERATO	
ENB-2	Digit span	Non somministrato	ALTERATO		
CRI	CRI-Totale	103	85-114	Medio	
	CRI-Scuola	98			
	CRI-Lavoro	102			
	CRI-Tempo Libero	106			
	ESAME NEUROCOMPORTAMENTALE	Non somministrato		ALTERATO	
WAIS IV	F. GREZZI	CUT OFF	ESITO		
Parole PZ	Non somministrato		ALTERATO		
ENPA	Ripetizione parole	Carzozzi 7/10	9/10	ALTERATO	
	Lettera parole	1/1	2	ALTERATO	
	Scrittura parole	3/10	0,3	6,4	ALTERATO
	Scrittura parole	1/1	1	4	ALTERATO
	Scrittura parole	1/1	0,9	1,3	ALTERATO
	Scrittura parole	3/10	3	8,2	ALTERATO
	Scrittura parole	4/10	3,5	6,1	ALTERATO
	Scrittura parole	18/20	18,4	18,4	LIMITI
	Scrittura parole	5/10	4,8	8,2	ALTERATO
	Scrittura parole	4/10	3,3	7,6	ALTERATO

CRI= 103

PATIENT 3 (FIRST ASSESSMENT)

GG	VALUTAZIONE ATTUALE: 14/10/11			
TEST	PUNT. GREZZO	CUT-OFF	ESITO	
ENB-2 e ALTRI TEST	Digit span	6/8	5	Norma
Test di Memoria di prosa - immediata	10/28	6	Norma	
Test di Memoria di prosa - differita	18/28	9	Norma	
Test di Memoria con interferenza - 10 s.	5/9	5	LIMITI	
Test di Memoria con interferenza - 30 s.	6/9	4	ALTERATO	
Trail making test - A	126"	66"	ALTERATO	
Trail making test - B	418"	149"	ALTERATO	
Test del gettoni	5/5	5	Norma	
Test di Elicitta fonemica	7/3	9	ALTERATO	
Test di Astrazione	6/6	4	Norma	
Test delle Figure Aggravate	5/5	4	Norma	
Test delle Figure Aggravate	16	28	ALTERATO	
Copia di disegno	2/2	2	ALTERATO	
Disegno spontaneo	2/2	2	Norma	
Test dell'Orologio	7/10	7	LIMITI	
Prove pratiche	6/6	6	Norma	
ENB-2 Punteggio Globale	60	73	ALTERATO	
CRI	CRI-TOTALE	125		Medio-alto
	CRI-Scuola	117		Medio-alto
	CRI-Lavoro	130		Medio-alto
	CRI-Tempo Libero	109		Medio
MMSSE	MMSSE totale grezzo	26/30		
	MMSSE corretto	23/30	24	ALTERATO

CRI= 125

PATIENT 2 (FIRST ASSESSMENT)

CN	VALUTAZIONE ATTUALE: 04/02/2011			
TEST	PUNT. GREZZO	CUT-OFF	ESITO	
ENB-2	Digit span	4/8	4	Norma
Test di Memoria di prosa - immediata	3/28	6	ALTERATO	
Test di Memoria di prosa - differita	8/28	9	ALTERATO	
Test di Memoria con interferenza - 10 s.	5/9	4	Norma	
Test di Memoria con interferenza - 30 s.	2/9	3	ALTERATO	
Trail making test - A	118"	56"	ALTERATO	
Trail making test - B	66"	214"	ALTERATO	
Test del gettoni	5/5	5	Norma	
Test di Elicitta fonemica	6/6	8	ALTERATO	
Test di Astrazione	4/6	3	Norma	
Test delle Figure Aggravate	5/5	4	Norma	
Test delle Figure Aggravate	23	25	ALTERATO	
Copia di disegno	2/2	2	Norma	
Disegno spontaneo	2/2	2	Norma	
Test dell'Orologio	8/10	6	Norma	
Prove pratiche	6/6	6	Norma	
ENB-2 Punteggio Globale per età	57	63	ALTERATO	
ENB-2 Punteggio Globale di Normalità	60	66	ALTERATO	
ALTRI TEST	Hamilton Depression Rating Scale	12		

CRI= 84

PATIENT 4 (FIRST ASSESSMENT)

M.E.	VALUTAZIONE ATTUALE: 08/11/2013			
TEST	PUNT. GREZZO	CUT-OFF	ESITO	
MMSSE	MMSSE totale grezzo	23/30	24	ALTERATO
ENB-2 e ALTRI TEST	Digit span	6/8	5	Norma
Test di Memoria con interferenza - 10 s.	5/9	3	Norma	
Test di Memoria con interferenza - 30 s.	6/9	3	Norma	
Test di Memoria con interferenza - 30 s.	6/9	3	Norma	
Test di Memoria con interferenza - 30 s.	6/9	3	Norma	
Test di Memoria con interferenza - 30 s.	6/9	3	Norma	
Test delle Figure Aggravate	23	27	ALTERATO	
FCST	IFR (Immediate Free Recall)	19 (17,12)	19,59	ALTERATO
	IFR (Immediate Free Recall)	32	35	ALTERATO
	DFR (Delayed Free Recall)	7	6,31	LIMITI
	DFR (Delayed Free Recall)	(6,70)	11	<11
	ISC (Index of Sensitivity of Cueing)	0,76	<0,9	ALTERATO
	n° intrasmi	6	8	ALTERATO
VIENNA TEST SYSTEM	WAFG - UNIMODALE - funzioni percettivo-attentive S2 (della allarme)	18 (RP 2)	<16	ALTERATO
	WAFG - CROSSMODALE - funzioni percettivo-attentive S2	n.e.		ALTERATO
	Schnep (interferenza lettura)	0,09 (RP 34)	<16	Norma
	Stoop (interferenza denominati)	0,08 (RP 46)	<16	Norma
	NIV - Nback verbale (reazioni sbagliate)	11 (RP 18)	<16	LIMITI

CRI= 137

RESULTS

In patients with low CRI the neuropsychological profile was clearly impaired at the first clinical assessment. Furthermore, significant cognitive deficits emerged at the psychometric tasks, confirming the findings of previous studies on the relationship between low cognitive reserve and cognitive profile. However, even in patients with high CRI, in which a good modulation of symptoms (i.e. compensation) is expected, test performances were clearly impaired, showing no protective effect of cognitive reserve.

DISCUSSION

High CR typically provides the preservation of cognitive functions in patients with a neurodegenerative disease, but it has weak or mild effects on YOD. Our data support Fairjones et al.'s findings (2011) which have showed that an early onset of Alzheimer's disease leads to a possible "erosion" of CR, particularly of memory.

CONCLUSION

In YOD general cognitive profile and memory function could only marginally benefit from the protective role of high CR. During any neuropsychological assessment, it is therefore fundamental to evaluate CR to give the symptoms the right interpretation, especially in young patients who might show only slight deficits.

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