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# Rorschach assessment of personality functioning in patients with polycystic lipomembranous osteodysplasia with sclerosing leukoencephalopathy

Ilonen T, Hakola P, Vanhanen M, Tiihonen J. Rorschach assessment of personality functioning in patients with polycystic lipomembranous osteodysplasia with sclerosing leukoencephalopathy.

**Objective:** To date no studies have investigated the personality functioning underlying patients diagnosed with polycystic lipomembranous osteodysplasia with sclerosing leukoencephalopathy (PLOSL) using the performance-based Rorschach test.

**Methods:** We scored and interpreted the Rorschach protocols of eight carefully diagnosed PLOSL patients according to Exner's Comprehensive System. The structural variables in the Rorschach are organised around the seven dimensions of personality functioning that they assess: coping style and resources, organising information, perceiving events, forming concepts and ideas, handling of emotions, self-perception and interpersonal perception.

**Results:** As a group PLOSL patients had many personality liabilities when contrasted with typical avoidant non-patient adults. A majority of patients showed an avoidant coping style (Lambda > 0.99), low productivity and poor verbal output in the low number of responses and few Blends. Also, they showed limited available resources to cope with problem-solving test. Problems in organising information efficiently, and perceiving events realistically as well as signs of disturbed thinking and concept formation were observed in many patients. Handling of emotions was characterised by avoidance of emotional stimuli but at the same time poor affect modulation. Capacities to view themselves and others were thoroughly limited.

**Conclusions:** PLOSL tends to have repercussions throughout the personality functioning. In line with clinical findings and later neuroradiological and neuropathological examinations the Rorschach revealed personality features typical for frontal type of dementia.

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# **Significant outcomes**

- PLOSL, which is a rare fatal disease affecting nervous and musculoskeletal systems, also deeply affects personality.
- Polycystic lipomembranous osteodysplasia with sclerosing leukoencephalopathy (PLOSL) patients show similar personality features to those typical for frontal type of dementia.
- The Rorschach, as a perceptual-cognitive problem-solving test, is suitable also for patients who are neurologically impaired.

# Limitations

- The data were collected mostly in third and fourth illness phases, thus we do not know the primary personality functioning of these patients.
- We were able to analyse only a part of the collected data because those in the most severe disease phase were not able to produce valid Rorschach protocols.
- Small sample size caused problems for statistical analysis.

## Introduction

PLOSL or Nasu-Hakola disease is a rare fatal disease affecting both brain and bones. This disease has been found in the 1960s in Finland and Japan (1,2). About 200 patients have been diagnosed in the world, most in Japan (about 100 cases) and in Finland (35 cases), but also in Sweden, Norway, USA, Italy and many other countries (3). PLOSL is a recessively inherited disease (1,4). The team of late Leena Peltonen-Palotie has first located the mutation on the chromosome 19q13.1 (5), then identified it as DAP12 gene in all Finnish and some Japanese patients (6,7). The mutation in 100% of Finnish patients and some Scandinavian patients is deletion of exons 1-4, in Japanese patient deletion of G 141 (3). Surprisingly, the mutation in TREM2 gene can cause clinically entirely similar disease picture as the mutations in the DAP12 (TYROBP) gene (8). Both DAP12 and TREM2 are expressed by central nervous system microglial cells and oligodendrocytes. DAP12 deficiency results in functional defects of these cells and also leads to synaptic abnormalities. Molecular genetic studies have also found that the mutations in the DAP12 gene cause disorders in the function of the osteoclast cells in the bones (9-11). Regrettably, these studies have not yet lead to any result helpful in the care of PLOSL patients.

The course of PLOSL is steady progressive. It has been divided into four stages: 0, 1, 2 and 3 (1,4). The childhood is mostly symptomless (the latent phase = 0) but around the age of 20 years patients start to suffer from pains and tenderness in the ankles and wrists and later from fractures (the phase of skeletal symptoms = 1). The radiological picture is diagnostic (1,12). The clinical neuropsychiatric features of PLOSL are known on the ground of, for example, a follow-up study of 22 PLOSL patients (4). Neuropsychiatric symptoms appear insidiously during the fourth decade of life and gradually form a disease picture which consists of dementia with prefrontal psychosyndrome, signs of upper motor neuron involvement apraxic-aphasic symptoms and myoclonic twitches (the early phase of neuropsychiatric symptoms = 2). These symptoms progress steadily and they are later accompanied with

other symptoms, especially epileptic seizures (the late phase = 3). The death follows before the age of 50 years.

The picture of the neuropathological changes in PLOSL has been well outlined. On neuropathological examination, atrophy and sclerosis of the white matter are found particularly in the frontal and temporal lobes of the brain (3,13–18). The changes in the brains of the patients still living are visible with the neuroradiological methods: PEG (19,20), MR and CT (21). Electroencephalogram shows also alterations (22).

In the early phase of the neuropsychiatric symptoms the most prominent symptoms are the frontal psychosyndrome and the advancing deterioration of memory. These both symptoms have been described *in casu* in the monographs of Hakola (1,4). The deterioration of visual memory has been published separately (23) as well as the defect of verbal memory (24). The test pattern used by Hakola in his follow-up studies contained the Rorschach test also to assess personality functioning of these patients. Some findings have been described formerly in these monographs (1,4) but as far as we know, any broader report about the results in the Rorschach test has not been formerly published in this fatal but fascinating disease.

Rorschach (25) found that the strategies people use to formulate inkblot responses allow descriptions of how they are likely to handle realworld situations. He was the first to examine responses of organically ill individuals to the inkblots. Later, Piotrowski (26) examined cortical and subcortical patients and introduced 10 signs of organic conditions. Dorken and Kral (27) found that patients with senile dementia have difficulties to adequately respond in the test. The number of responses was low, and thus they offered only few movement, colour and shading determinants. Poor form quality was related to their level of deterioration. In a study of Insua and Loza (28), the lack of percept involving human movement, indicating a weak energy level, differentiated the elderly demented group from the normal comparison group. Perry et al. (29) studied patients with dementia of the Alzheimer type. They found

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significant differences between Alzheimer patients and normal controls for most of the Rorschach test variables.

PLOSL patients are characterised by frontal type of dementia. Frontal lobe disturbances tend to have repercussions throughout the behaviour (30). According to Lezak (31), the behavioural disturbances with frontal lobe damage can be classified into problems of starting (e.g. decreased productivity), difficulties in making mental or behavioural shifts (e.g. perseveration, rigidity), problems in stopping (e.g. impulsivity, loss of control), deficient selfawareness (e.g. inability to perceive events) and a concrete attitude. Patients with frontal deficits are expected to produce an inefficient approach to the task. There are many relevant Rorschach variables that relate to behaviour described in patients with frontal lobe disturbances. On the basis of the earlier studies of neurological disorders and the behavioural disturbances with frontal lobe damage described by Lezak (31), we hypothesised that in comparison to normative data (32) PLOSL patients have behavioural disturbances.

# Aims of the study

The purpose of this study was to describe personality functioning of PLOSL patients with the Rorschach test. The structural variables in the Rorschach are organised around the seven dimensions of personality functioning that they assess. We investigated PLOSL patients' coping style and resources, how they organise information, how they perceive their events, how they form concepts and ideas, how they handle their emotions, how they view themselves and how they relate to other people.

# **Materials and methods**

Subjects

Material comprised 21 of the 35 PLOSL patients diagnosed until today in Finland (Table 1). Our 21 patients included all cases that could be reached for evaluation, and 14 patients were excluded due to practical reasons such as remote geographical location. Therefore selection was random and not by the severity of the illness. They were 11 male and 10 female. The mean age was 34.9 (SD = 4.3; range = 28–45). In all cases, the diagnosis was confirmed with radiographs of bones. In addition to this, typical neuropsychiatric symptoms were observed in clinical examinations (in one case after Rorschach test), and in most cases, typical findings were observed also in neuroradiological examination.

Rorschach assessment

All patients were administered the Rorschach test by Dr Hakola (n = 14), or psychologists Luostarinen (n = 3) or Peltokorpi (n = 4). The protocols were scored and interpreted following Exner's (32,33) Comprehensive System guidelines with the structural summary produced through the Rorschach Interpretation Assistance Program (version 5) by the first author who is an expert in using the method. Reliability and validity estimates are established and described extensively elsewhere (32-35). The protocols with more than 13 responses (R) provide reliable data and support valid inferences (33). Thirteen of the 21 patients could not respond to the test sufficiently, and thus we included only eight valid protocols (R > 14) for analysis. The mean age of patients with R > 14 was 32.1 (SD = 2.6; range = 28-36). Cases 1, 2, 3, 4, 6, 9, 14 and 16 produced a valid protocol for interpretation (Table 1). All but one (case 4) was on disease phase 1 or 2. The symbols in Table 1 refer generally to the time when the Rorschach test was performed. Only exceptions are the epileptic seizures which have begun at the time presented, and the brain atrophy in neuroradiological and/or neuropathological examination considerably

The selected personality variables and interpretive cut-off scores as presented by Exner (32) are presented in Tables 2 and 3.

## **Results**

The results of the Rorschach assessments are summarised in Tables 2 and 3. PLOSL patients gave responses for 11 variables (R, Lambda, DQ+, PSV, XA%, WDA%, P. X-%, WSum6, Lv2 and PureC) that were significantly different from normative sample of avoidant adults with z-scores more than 1 SD from the mean (Table 2). As shown in Table 3, using interpretive cut-off scores as presented by Exner (32), a majority of PLOSL patients showed an avoidant coping style, low productivity, poor verbal output and limited resources to cope with problem-solving task. Half of the patients demonstrated abstraction difficulties in low number of movement responses, and more than half low level of emotional stress experience. Problems in organising information efficiently, and perceiving events realistically as well as signs of disturbed thinking and concept formation were observed in many patients. Handling of emotions was characterised by avoidance of emotional stimuli but at the same time poor affect modulation. Low self-esteem was observed in half of PLOSL patients. Capacity for being introspective was limited in almost all of PLOSL patients. Half of the patients scored in the pathological range

Table 1. The patients by sex, age and most important neuropsychiatric, neuroradiological and neuropathologiacal findings

Case	*	2*	3*	4*	2	*9	7	8	*6	10	11	12	13	14*	15	16*	17	18	19	20
Sex	Σ	Σ	ч	Σ	т	щ	ч	ч	щ	Σ	ч	Σ	ч	Σ	ч		4	Σ		⋝
Age	35	31	30	36	35	32	38	45	33	31	37	34	42	28	40	32	31	40	34	38
Disease phase	2	2	2	က	2	2	က	က	2	က	က	က	က	_	က		3	က		က
Prefrontal psychosyndrome																				
Euphoria	+	+	+	+	+	+	+		+	+		+	+		+	+		$^{\rm H}$		
Witzelsucht	+	+	+	+	+	+	+		+	+		+	+		+		+			
Lack of insight	+	+	+	+	+	+	+	+	+	+	+	+	+		+	+		+	+	Н
Lack of self-control			+	+	+	+	+	+		+	+	+			+	+				
Lack of social inhibitions		+	+	+	+	+	+		+	+	+	+	+		+					
Inclination to immediate reactions		+		+	+	+	+			+			+		+	+				
Impaired capacity to concentrate		+	+	+	+	+	+	+	+	+	+	+			+			+	+	+
Lack of initiative				+	+	+	+	+	+	+	+				+	H		H		
Deterioration of memory																				
Visual	+	+	++	++++	+	+	+ + +	+ + +	‡	+ + +	++++	+	+	•	++++		_	++++	+	+
Verbal	+	++	+	++		+	++	+++	+	++	++++	++	++		++	+	+ ++	+ + -		+
Other neuropsychiatric symptoms																				
Aphasia		+	+	+	+	+	+	+		+	+		+				+1			
Apraxia			+	+			+	+	+	+	+		+					+		
Signs of I motor neuron involvement	+	+	++	++	++	+	+	++	+	+	++	+			+	+	+	++		+
Epileptic seizures		+	+	+	+	+	+	+	+	+	+				+				+	
Age at the first epileptic seizure		41	32	41	42	37	37	46	į	25	34				39				43	
Brain atrophy																				
In neuroradiological examination	+ :	+ -	+ - + - + - + - + - + - + - + - + - + -	+ -	+ :	+ + :	+ + : +	+ + +	+ + +	+ :	+	+ + +	+	+	+	<b>+</b>		+	+	
ın neuropatnological examination	+	+++++	+	+++++	+	+	+			+ + +										

\*Produced valid Rorschach protocol. Disease phase 1 = mild, 2 = marked, 3 = severe deviation. Prefrontal psychosyndrome + = symptom existed, ± = uncertain. Deterioration of memory, other psychiatric symptoms and brain atrophy + = mild, ++ = moderate, + + + = severe. Case 14 showed no neuropsychological symptoms at the time when Rorschach was done. However, he developed florid symptoms during follow-up afterwards.

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Table 2. Descriptive statistics for selected Rorschach variables from PLOSL patients and normative sample of avoidant adults (32)

		PLOSL,	n = 8	Normative avoidant adu		
Variable	Descriptive	Mean	SD	Mean	SD	Z
R	Number of responses	17.25	4.9	22.83	5.3	-1.05
Lambda	Openness to experience (avoidant if Lambda > 0.99)	2.21	2.0	1.33	0.4	2.20
Blends	More than one determinant in explaining the percept	1.50	1.3	3.07	1.7	-0.92
EA	A combined ideational/affective index of resources available	4.18	1.9	5.93	1.9	-0.92
M	Responses in which people or animals are engaged in human activities	1.37	0.9	2.83	1.5	-0.97
FM+m	Animal movement + inanimate movement	2.25	1.6	3.53	2.1	-0.61
SumShading	An index of stress that is experienced as a consequence of painful emotions	1.38	1.59	2.33	1.8	-0.53
D-score	Capacity for control and tolerance for stress	0.12	1.0	-0.07	0.9	0.05
AdjD	Capacity for control and tolerance for stress	0.25	0.7	0.02	0.8	0.29
Zd	Efficiency in organising information	1.50	3.3	-1.12	2.7	0.14
DQ+	Two or more separate objects are seen as related to each other	3.38	3.1	5.55	1.7	-1.28
DQv	An object having no specific form is identified	1.38	1.1	0.91	1.6	0.29
PSV	Perseveration	1.75	2.1	0.19	0.4	3.90
XA%	Appropriate use of form features	60.00	12.8	91.0	10.0	-3.10
WDA%	Appropriate use of form features in common areas	62.75	16.0	92.0	10.0	-2.93
Р	Popular responses	3.88	2.0	6.31	1.2	-2.03
X-%	Distorted form level responses	36.12	14.8	9.00	10.0	3.60
WSum6	The sum of weighted values of the special scores for unusual forms of expression	14.62	17.4	5.21	3.3	2.85
Lv2	Severe cognitive disruption	0.87	1.5	0.07	0.3	2.67
WSumC	Weighted sum of colour responses	2.81	1.5	3.10	1.4	-0.21
Afr	Affective ratio; an index of receptivity to emotional stimulation	0.39	0.1	0.56	0.2	-0.85
PureC	Pure colour responses	0.88	0.6	0.12	0.4	1.90
3r+(2)/R	Egocentricity index	0.33	0.2	0.40	0.1	-0.70
MOR	Morbid content	2.25	2.3	0.62	0.6	2.71
V	Self-critical attitude	0.12	0.4	0.12	0.5	0
FD	Capacity for being introspective	0.12	0.4	0.57	0.7	-0.64
SumH	Sum of human responses; interpersonal interest variable	3.50	2.4	4.97	1.5	-0.98
M+,Mo,Mu	Adequate empathic capacity	1.25	0.9	2.80	1.9	-0.82

on the coping deficit index (CDI), indicating a lack of social skills and ineffective ways of attempting to cope with ordinary aspects of everyday living. More than half of the patients seemed to have a normal interest in other people but empathic capacity was inadequate.

In 13 cases, the protocols contained less than 14 responses (R ranged 8-13), and thus these were not possible to analyse using Exner's (32,33) Comprehensive System guidelines. The majority of these patients suffered already the late phase of PLOSL (10/13), and they were older (mean 36.7 + 4.3) than the patients of the group analysed (mean 32.1 + 2.6).

## Discussion

This study is the first to describe personality functioning in patients with PLOSL. As expected the overall approach to the test was constricted. Our results are in line with previous studies that have demonstrated that patients with neurological disorders perform differently than healthy controls on the Rorschach test (26–29). PLOSL patients showed more or less

difficulties on all seven dimensions of personality functioning. As potential psychological indices of prefrontal impairment the patients showed low productivity in the number of responses, and consistently a decrement in verbal abilities in low number of Blends.

What kind of coping style characterises PLOSL patients? Can they muster adequate resources for meeting the demands they are facing? Successful adaptation requires openness to experience and mustering adequate resources. In our study, all but one patient who was ambitent, showed an avoidant coping style. Avoidant style includes a marked disposition to simplify complexity by disregarding or even denying some aspect of stimulus event (32). Ambitents are people who do not have a consistent approach to problem solving. The lack of consistency in the use of feelings may cause ambitents to become confused by them or lead to erratic forms of emotional display (PureC). On the other hand, the lack of consistency in the manner by which they conceptualise and make decisions reduces efficiency and leads to errors in judgment. Because of the inconsistency and lack of flexibility of their

Table 3. Personality functioning in the PLOSL patients

Variables	Ν	%
Coping style and resources		
Avoidant (Lambda > 0.99)	7	87.5
Low productivity (R < 17)	6	75.0
Poor verbal output (Blends < 4)	8	100.0
Low access to resources (EA $< 6.5$ )	7	87.5
Low stress tolerance (D $<$ 0; AdjD $<$ 0)	1	12.5
Abstraction difficulties (M < 2)	5	62.5
Mental activity provoked by need [(FM $+$ m) $<$ 3]	5	62.5
Low level of emotional stress (Sum Shading < 5)	7	87.5
Organising information		
Underincorporators (Zd $< -3.0$ )	1	12.5
Overincorporators (Zd $> +3.0$ )	4	50.0
Immature perceptual organisation (DQ+ $<$ 5 and DQv $>$ 0	6	75.0
Increase in perseveration (PSV $> 0$ )	5	62.5
Perceiving events		
Unconventional reality perception:		
(XA% < 70)	6	75.0
(WDA% < 75)	5	62.5
Popular responses ( $P < 4$ )	4	50.0
Distorted form perception (X-% > 20)	7	87.5
Forming concepts and ideas		
Disturbed thinking (WSum6 > 12)	3	37.5
Lv 2 disturbance	3	37.5
Handling of emotions		
Depression index, DEPI > 5	3	37.5
Restricted expression of emotions (WSumC < 2.5)	3	37.5
Avoidance of emotional stimuli (Afr < 40)	5	62.5
Poor affect modulation (PureC)	6	75.0
Painful affects (Sum Shading > FM+m)	2	25.0
Self-perception		
Low self-esteem $[3r+(2)/R < .33]$	4	50.0
Morbid content (MOR >2)	3	37.5
Poor self-introspection ( $V = 0$ or $FD = 0$ )	7	87.5
Interpersonal perception		
Social incompetence (CDI >4)	4	50.0
Low interest in other people (SumH <4)	3	37.5
Low empathic capacity (M+, Mo and Mu $<$ 2)	6	75.0

Rorschach variables and interpretive cut-off scores used in this study based on Exner (32).

coping style PLOSL patients show high vulnerability to experience adjustment problems. In addition, they showed fewer resources available than most people for coping with the ordinary ideational and emotional everyday life demands. Low number of human movement responses indicates limitation of the ability for higher order cognitive elaboration. Still most of the patients had D and AdjD scores in the zero or plus range indicating adequate frustration tolerance and adaptive capacity for stress control. However, along with avoidant coping style and fewer resources available stress tolerance is likely to be overestimated. These patients were poorly responsive to environmental demands. The attention paid to internal stimuli also was poor. Because of their limited selfawareness PLOSL patients are at risk for becoming precipitated into limited frustration tolerance, and poor impulse control, typical for frontal lobe damage (31).

How carefully and thoroughly PLOSL patients pay attention to events in their lives, and how they organise their perceptions? Organising information efficiently consists of an adaptive balance between the amount of information individuals take in and their capacities to process this information adequately. Successful adaptation requires efficient organisation of information. Processing efficiency variable Zd ranges normatively from -3.0 to +3.0. Within this range individuals tend to be taking in just as much information as they can process adequately. In our study one patient was underincorporator, i.e. he/she took in too little information. Thus he/she did not pay attention to relevant considerations but came to conclusions hastily. By contrast, half of the patients were overincorporators taking in more information than most people require in order to make decision. These patients invested effort and energy into scanning activities, but despite having made their best efforts they at the same time were hesitant and uncertain in making decisions. Further, DQ distribution refers to the quality of the processing activity. Low DO+ and high DQv showed that the quality of their perceptual organisation is immature and regressive. Thus, their cognitive activity is concrete and impressionistic representing a diffuse and inept form of processing typical for neurologically impaired persons (32). Also the presence of perseveration responses suggests problems in processing efficiency reflecting cognitive inflexibility and incapacity. Perseveration is common finding in individuals who have neurologically related problems (32). Let it be noted that the Rorschach test revealed tendency to perseverate earlier than the feature was seen clinically. Our findings are in line with earlier findings that patients with prefrontal damage show information-processing deficits that reduce their sensitivity to novel stimuli (36).

How PLOSL patients perceive their events? Successful adaptation is promoted by realistic and conventional perception of events and environments. This ability was measured by three form quality variables, Form Appropriate (XA%), Form Appropriate-Common Areas (WDA%) and Distorted Form (X-%). Perceptual disturbance is typically considered a primary feature of schizophrenia but is also observed in patients at high risk of psychosis (37). A majority of PLOSL patients demonstrated a severe impairment, often misperceiving events and forming mistaken impressions of people and what their actions signify. Their inaccurate perceptions of people and events are likely to lead them to erroneous conclusions and actions. Most people with this degree of impairment reality testing have difficulty in managing basic psychological aspects of everyday living without assistance.

How PLOSL patients form concepts and ideas? Forming concepts and ideas consists of how people think about the experiences they have and the impressions they form of events in their lives. People adapt best when they can think logically and coherently. Contrary to Perry et al. (29) who studied patients with dementia of Alzheimer type, signs of disturbed ideation were observed in some of PLOSL patients indicating abstraction and linguistic difficulties. Thus, some of PLOSL patients have lost capacity to think logically, and as a consequence tend to reason arbitrarily. Their answers including deviant verbalization and deviant response indicated that their ideas tend to emerge out of sequence or intrude on each other to produce rambling or tangential and unintelligible verbalisations.

How PLOSL patients experience and express feelings? Handling of emotions provides information about the manner with which people process emotional experience. Three patients (37.5%) had depression index (DEPI) = 5 indicating that their psychological organisation was marked by features that give rise to experiences of emotional disruption. One of those whose DEPI was positive also CDI was positive indicating depressive features mainly characterised by hopelessness and helplessness because of interpersonal difficulties or social incompetence. Differently from most depressive individuals they did not demonstrate data of sad feelings or anger although they had to know they were close to death. Their ability to recognise their inner state and feelings was limited. Perhaps the lack of sadness made it difficult to identify clinically notable depression. Withdrawal from affective experience increased towards the most severe disease phase. Personality tended to fade. Consistently with the avoidant coping style, 37.5% of patients scored WSumC < 2.5 indicating insufficient capacity to experience and express feelings. On the other hand, PureC > 0 indicated impulsive affects and difficulties in affect modulation. The inability to modulate emotions and limited awareness of subjective feelings of distress [(FM+m) + Sum Shading = 3.6 + 2.3] may contribute to impulsive behaviour. Impaired emotional processing and deficits in the ability to recognise the affective state of other people characterise also patients with Alzheimer's disease (38). Disordered expression of affect is frequently a result of frontal system lesions (30).

How PLOSL patients view themselves and how they relate to other people? Capacities to view

oneself in favourable ways maintain adequate selfesteem and contribute to a stable sense of identity. Now, low self-esteem was found in half of PLOSL patients indicating that they tend to judge themselves more unfavourably in comparison with others. Further, MOR answers reflect either pessimistic thinking or signal that their self-image is marked by many negative attributions in which they tend to perceive themselves as damaged or distorted. The absence of introspection suggests that they are also rather naïve about themselves. They do not have a grasp of the kind of persons they are. Lack of selfawareness may be a risk for adjustment difficulties. Normally, the abilities to sustain a reasonable level of interest in interacting with other people and to perceive people and social situations in an accurate and empathic manner characterise adaptive interpersonal relationships. Disorders of social behaviour may suggest frontal lobe dysfunction (39). In interacting with others, PLOSL patients are characterised by social incompetence (50% of patients with CDI > 3), low empathic capacity and limited interest in people. Their social ineptness may make them vulnerable to experiencing failure in social situations and to being ignored by others who see them as an ineffective person (32).

The findings of our study provide the first descriptive statistics for the Rorschach test in patients with a rare fatal neurological disease. One obvious problem with our sample was a small sample size. Those who were in the most severe disease phase were not able to produce valid Rorschach protocols evidencing that impairment in personality functioning becomes more pronounced and worsens with illness progression. Further, our study was cross-sectional and patients were tested during phases 2 and 3. In future it is important to assess personality functioning across all different illness phases to identify changes in personality dimensions. It was interesting that as a perceptual-cognitive problem-solving task, the Rorschach test was able to use with patients who are neurologically impaired and even intellectually limited. Our study may shed further light on the multifaceted clinical phenomenology and psychopathology of PLOSL. The personality profile of PLOSL patients resembles that of frontal personality disturbance, and is in line with clinical findings. Our results are consistent with study of Stuss et al. (30) who reported that frontal lobe disturbances tend to have repercussions throughout the behaviour.

In summary, PLOSL patients have difficulty attending to their surroundings openly, efficiently, realistically and conventionally. Thinking tends to be disturbed, affect modulation insufficient and

capacities to view themselves and others thoroughly limited.

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