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Integrated cognitive retraining in agenesis of corpus callosum: a single case study

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ABSTRACT

Individuals with Agenesis of corpus callosum (ACC) have deficits in areas of cognitive functions, social, emotional processing and communication. The aims of the study were to examine: (a) the neuropsychological profile pre- and post-intervention; (b) to study the effects of the integrated integrative cognitive retraining in patient with ACC. We employed a single case experimental study with pre- and post-assessment. This design enabled evaluation of changes in the patient in response to the intervention. RR, a 25-year-old with diagnosis of ACC was assessed for poor concentration, memory, academic performance, anxiety, and interpersonal issues. Neuropsychological profile indicated deficits. He underwent six months of cognitive retraining (home & hospital based) and brief cognitive behavior therapy. There was significant improvement post intervention. Integrative Cognitive retraining was efficacious in patient with ACC.

Keywords: Agenesis of Corpus Callosum; Integrative Cognitive Retraining; Neuropsychological Assessment

1. INTRODUCTION

Agenesis of the corpus callosum is among the most common brain malformation observed in humans (Dobyns, 1996). ACC is failure to develop the large bundle of fibres that connect the cerebral hemispheres, occurs in 1:4000 individuals. Studies using neuropsychological, electroencephalogram and functional MRI approaches are examining the resulting impairments in emotional and social functioning, and have begun to explore the functional

neuroanatomy underlying impaired higher-order cognition. (Paul et. al., 2007). The clinical correlates of ACC are associated with learning difficulty and epilepsy (Aicardi, Chevrie & Barnton, 1987) and with neuropsychiatric disorders, including attention-deficit hyperactivity disorder (Hynd, Hall & Novey, 1995), schizophrenia (Lewis, Reveley & David, 1988). In a study by Paul & Brown (2000) it was found that individuals with ACC have normal-range IQs. They also found that deficits in complex cognition manifested in psychosocial functioning. In terms of language patients, tend to offer meaningless or out-of-place comments in conversation, they have difficulty understanding the subtle aspects of social context, and have difficulty comprehending second order meanings in language (e.g., in jokes, stories, and non literal expressions). The other forms of language deficits in patients with ACC were impairment in phonemic discrimination, phonological reading (Temple et al., 1989; 1990), processing and use of nonliteral language (Brown et al., 2005; Paul et al., 2003). With regard to difficulties in social processing researchers have hypothesized that the social deficits were secondary to impairments in novel complex problem solving (Brown & Paul, 2000). Studies with ACC also show impairment in the problem solving in both non-emotional tasks (Brown & Paul, 2000; Garrels et al., 2001; Schieffer et al., 2000; Symington et al., 2004) and complex tasks involving emotional stimuli (Paul et al., 2004; Symington et al., 2004). Complex spatial memory deficits (Temple & Ilsley, 1994), deficits in concept formation and problem solving (Schieffer et al., 2000), and difficulties in their ability to learn from feedback, strategize, and imagine consequences for their actions (Symington et al., 2004) were also reported. Poor social insight and peer relationships (Brown & Paul, 2000), poor social problem solving and social judgment (Symington et al., 2004) have been reported. The impaired theory of mind has been proposed by Symington et al., in 2004. There are indications that and an inadequate understanding of pictures portraying complex social interactions in individuals with ACC (Paul et al., 2004; Turk et al., 2003). Research show that patients with ACC show deficits on cognitive emotional and social aspects. This study is aimed to examine the neuropsychological profile of a patient. The studies with regard to retraining of these neuropsychological deficits have been scanty. Hence this study is an attempt to evaluate cognitive retraining in ACC.

2. METHOD

A single case experimental study with pre- and post-assessment was used. This design enabled evaluation of changes in the patient in response to the intervention.

2.1. Participant back ground information

RR, a 25-year-old, right-handed male, student of a MBBS course, hailing from upper middle class, urban background was diagnosed with agenesis of corpus collusum in 2000. He was started on sizodon and cytopam medication for 3 years. He developed mild sizodon induced parkinsonisium. A MRI scan showed complete agenesis of corpus collosum in December 2005. He also sustained a severe head injury in a car accident January 2005 (GCS 8/15) with loss of conscious for 24 hours, with 3 days of altered sensorium. CT scan in June 2005 showed mild subarachnoid hemorrhage with a cerebral atrophy. Since 2000 his complaints were poor concentration, poor memory, anxiety symptoms and poor academic performance. His performance deteriorated to the extent of inability to clear second semester of the course, even after attempting 6 times. Personal history suggests that he had poor interpersonal relationship with father. Temperamentally he was described to be shy, sensitive and introvert. Father was over involved and was critical towards patient. RR underwent neuropsychological assessment in January 2006.

2.2. Procedure

With informed consent the patient was assessed with adequate rest pause during the testing. The patient's pre-training neuropsychological assessment revealed impairment in motor speed, verbal learning and memory and visual learning and memory. The rational for Home Based Cognitive Retraining HBCR (Rao et al., 2004) was due to patient's inability to stay in the hospital for treatment. The Home based tasks was chosen to ameliorate the deficits in a graded manner, according to the following schedule. Letter cancellation was used to improve attention. Information processing was enhanced with grain sorting. Emphasizing careful regulation of pressure and direction of strokes as well as neatness increased response inhibition with coloring task. Jumbled sentences, mental arithmetic, visuo-spatial encoding and temporal encoding was used in addition to improve working memory, visual and verbal memory. Hospital Based Retraining included the same task for one month. The computer tasks were introduced. The tasks included joining the dot, coloring, puzzles, and jumbled letters, mazes. The computer based items were predominantly to enhance his executive functions. These functions were targeted in weekly. The cognitive retraining was carried out daily at home by the patient for 1 ½ to 2 hours by father. Feedback on the performance of the retraining tasks was taken from the father and the patient at the end of each week. Based on saturation cue technique tasks were gradually increased

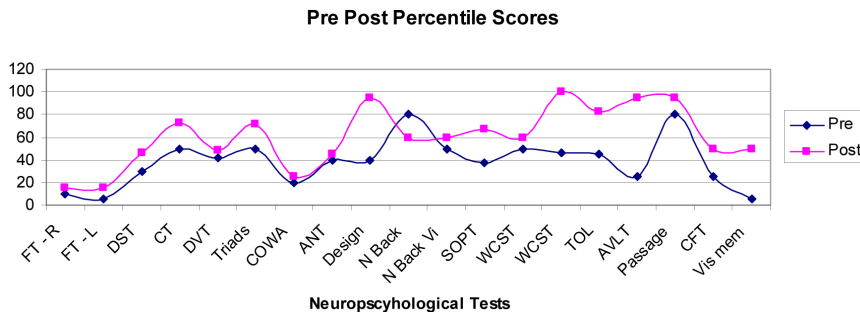
in the level of difficulty. Totally the HBCR and Hospital based retraining was carried out for 6 months. After the completion of cognitive retraining a post Neuropsychological assessment was carried out. He was suggested to use diary as an external aid. During the course of treatment patient's anxiety in the interpersonal situation, academic performance, and self esteem issues emerged. Hence the component of cognitive restructuring was included to elevate his anxiety symptoms and fear of negative evaluation. The individual sessions focused on relaxation techniques and cognitive restructuring. In relation to the fathers criticality, sessions, focused on assertive skills. Father was psycho- educated about patient's illness in terms of cognitive deficits and psycho social aspects.

3. RESULTS

Data Analysis: In order to examine the prevalence of cognitive deficits, a cognitive profile of the patient was compared with the normative data. Cognitive functioning of the patient was compared with normative data derived from a group of 540 normal healthy volunteers (Rao et al., 2004). Healthy volunteers were recruited from relatives of patients admitted at the hospital, students, and from the community at large. Healthy volunteers who obtained a score of > 2 on the General Health Questionnaire (GHQ-12) were excluded, as these persons met criteria for psychological distress, which could influence cognitive functioning. Healthy volunteers were also excluded if they had a previous history of neurological, neurosurgical, or psychiatric illness or substance dependence and family history of alcohol dependence, schizophrenia or bipolar disorder. For each test variable, percentile scores were calculated. The 15th percentile score (1 SD below the mean) was taken as the cut off score. Cutoff scores were calculated for each group based on age, education, and gender. A deficit was defined as a test score falling below the 15th percentile. Validation for each of the tests has been carried out on patient groups with focal lesions, refractory epilepsy, head injury, and Parkinson disease. In order to examine the prevalence of cognitive deficits, a cognitive profile of the patient was compared with the normative data described above. Based on the number of test variables falling below the 15th percentile, the severity of cognitive impairment was established. The percentile scores are given in the table and percentile scores are depicted in the graph.

Table 1. Percentile scores pre-post intervention of RR

NEUROPSYCHOLOGICAL ASSESSMENT Tests Administered	PERFORMANCE IN PERCENTILES	
	Pre	Post
Motor speed - Finger Tapping (FT) - Right	10 th p	15 th p
Finger Tapping - Left	5 th p	15 th p
Mental Speed - Digit Symbol Substitution Test (DSST)	30 th p	46 th p
Focused Attention - Color Trails (CT)	50 th p	73 rd p
Sustained Attention - Digit Vigilance Test Time (DVT)	42 nd p	48 th p
Divided Attention Triads Total Number Error	50 th p	72 nd p
Fluency		
Controlled Oral Word Association Test (COWA)	20 th p	25 th p
Category Fluency - Animal Name Testing (ANT)	40 th p	45 th p
Design Fluency	40 th p	30 th p - 95 th p
Verbal Working Memory Verbal N-Back Test	60-80 th p	20-60 th p
Visual Working Memory Visual N-Back Test	30-50 th p	20-60 th p
Self Ordered Pointing test (SOPT) Word Error	37 th p	67 th p
Set Shifting Wisconsin Card Sorting Test (WCST)	50 th p	60 th p
Failure to Maintain Set	46 th p	100 th p
Planning: Tower of London (TOL) - Mean Time	45 th p	83 rd p
Verbal Learning & Memory Auditory Verbal Learning Test (AVLT) - Trail	10-20 th p	50-75 th p
Immediate recall	25 th p	50-60 th p
Delayed recall	15-25 th p	85-95 th p
Passage Test - Logical Memory - Delayed Recall	80 th p	95 th p
Visual Learning & Memory - Complex Figure Test (CFT) - Delayed Recall	25 th p	50 th p
Design Learning Test Trial 1 Score	5 th p	50 th p
Design Learning Test Trial 4 Score	60 th p	85 th p



Finger Tapping Right (FT-R) Finger Tapping Left (FT-L), Digit Symbol Substitution Test (DSST), Color trials (CT), Triads Test, Controlled Word Association test, Anima Name Test, Design fluency, N-Back test (verbal & visual), Self Ordered Pointing Test (SOPT), Wisconsin Card Sorting Test (WCST), Tower of London (TOL), Auditory Verbal Learning Test (AVLT), Passage Test, Complex Figure Test (CFT), Visual Learning Test.

Figure 1. Percentile scores of neuropsychological functioning pre- and post-intervention

4. DISCUSSION

Single case study was used in which a patient with ACC and mild head-injury was compared pre and post neuropsychological profile. Patient underwent five months home based and one month hospital based cognitive retraining, which included attention/concentration, relaxation, executive functions, memory, cognitive restructuring and parental counseling. Prior to the intervention, patients had deficits in motor speed, verbal and visual learning and, memory. The assessment following the intervention showed significant improvement in the cognitive functions that were initially assessed to be in the deficit range. Cognitive rehabilitation techniques consist of three broad categories: restore, substitute, or restructure (Stephanie, 2002). The techniques for restoring RR's cognitive functions included cognitive training and exercises which was directed toward strengthening and the reestablishment of impaired abilities. He was suggested to use substitute techniques such as diary as an external devices which was meant to be a compensatory strategy to overcome memory difficulties. The session with individual and with father aimed at psycho social restructuring to reorganize his environment in order to compensate for impairment. The model which supports the recovery is through mechanisms of plasticity of the brain (Ricardo et al., 1997). According to Jennet, recovery after several days is more likely to be due to

resolution of temporary structural abnormalities, (1990), or to the depression of metabolic enzyme activity (Whyte, 1990). Kolb and Whishaw (2000) outlined three ways in which recovery is possible following brain injury. First, endogenous recovery may occur. Intact neural circuits may reorganize to accommodate functions that previously resided in damaged neural areas by dendritic proliferation. Second, recovery may have an exogenous origin in that cerebral reorganization may be prompted or accelerated by therapeutic interventions such as pharmacological agents or behavioral therapy. Third, recovery may not involve reorganization at all, but instead involve the replacement of injured neurons. The process of recovery after months or years is even less well understood. The cognitive retraining emphasis was also on psycho social factors which played a major role. As patients are not simply a manifestation of underlying cognitive problems, but are complex organisms within social and psychic environments (Wilson, 1997), the training incorporated techniques to enhance his emotional and psychosocial problems.

The present study concludes that the cognitive retraining was found to be useful for patient with ACC. On a visual analog scale of ten, patient and father report 8-9 point improvement in the over all functioning of the patient. The follow of up status of the patient was obtained from telephone conversation and email. He gradually cleared his exams by taking two or three subjects at a time within a span of 6 months, which was pending for almost 3 years. Finally patient cleared all his subjects and he was posted for internship. Overall along with improvement of neuropsychological performance, his self-confidence and self-esteem was reported to be enhanced, though there was no standardized scale measure. The study suggests usefulness of cognitive retraining in patient with ACC. The finding of the study suggests that more intensive research efforts in terms of outcome measures are needed in the area to validate long term effects. To conclude the study puts forward that cognitive retraining should be taken into account to plan for rehabilitation of patients with ACC.

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