



Bipolar Disorder in an Individual with Severe Developmental Disability: A Case Report

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JCPNP 2004;3(4).

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Peer Reviewed by two reviewers.

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Abstract

Due to unique diagnostic and treatment issues, many distinctive clinical situations are encountered in treating psychiatric disorders in those with severe-profound developmental disability and co-morbid psychopathology. This report reviews the effects of poly-pharmacotherapy on the measurable behavioral symptoms displayed by a person with severe mental retardation and an eventual diagnosis of rapid cycling bipolar disorder. The limited verbal skills entailed in this population require a different approach to diagnosis due to lack of useful verbal information exchange. Other obstacles encountered in arriving at a diagnosis and developing an effective treatment plan, including the matter of diagnostic overshadowing, are reviewed.

In the present case, clinically significant improvements in rates of measured behaviors including sleep patterns were observed after the discontinuation of paroxetine and mesoridazine, followed by the initiation of a lithium, olanzapine, and divalproex medication regimen. The patient's observed quality of life and functional ability improved with improved mood stability, subsequent to the above noted changes.

Conclusions: The importance of reviewing the current medication regimen as a potential contributor to the presenting problem is imperative. A detailed psychopharmacologic medication history is critical to facilitate sound treatment decisions. The importance of clinical perseverance via a well organized, carefully planned treatment approach is of vital importance.

Index terms: bipolar disorder, developmental disability, diagnosis

Introduction

Clinicians who do not specialize in treating individuals with mental retardation and comorbid psychopathology are often nonetheless faced with the unique diagnostic and treatment issues presented by this population. It is now widely accepted that individuals with developmental disability can manifest any psychiatric disorder seen in the non-developmentally disabled.^{1,2} However, psychiatric diagnosis and treatment in this population is replete with potential pitfalls, including under-recognition of the disorders.³ Another particular conundrum is correctly identifying the behavioral manifestations of a psychiatric disorder in those with severe to profound developmental disability, and thus limited verbal abilities.^{2,3,4}

One common mistake is that of attributing the observed behavior to an aspect of the mental retardation, commonly referred to as diagnostic overshadowing. This phenomenon is a common tendency among mental health professionals in which psychopathology is mis-identified as a learned behavior or as a facet of the mental retardation, thus failing to appreciate it as a potential component of a mental illness.^{3,5,6} Excellent reviews regarding the diagnostic issues related to mood disorders in those with severe-profound mental retardation have been published, with a central theme being that of diagnostic overshadowing.^{3,5,6}

Diagnosis of psychiatric disorders becomes increasingly difficult for individuals with severe-profound mental retardation due to verbal communication deficits. Language skills deficits significantly limit application of many diagnostic criteria, which are based upon patient interview and verbal interaction. Also, standard psychometric assessment tools are not designed for use in

individuals with severe to profound developmental disabilities. In addition, the actual clinical manifestation of a given psychiatric symptom may be altered in an individual with significant cognitive skills deficits.⁵ In such cases where a standard DSM-IV diagnosis cannot be made with confidence, treatment decisions may be based upon identification of specific, well defined, and measurable target behaviors. Clinicians are faced with making a "best fit" diagnosis, given the interpretation of the available information from observations and staff reports.⁶ A limitation of this method, however, is that such target behaviors may be associated with a variety of psychiatric disorders, and specific treatment may vary.⁵ This leads to a situation where clinical hypothesis testing is utilized (i.e. a series of planned medication and behavior support plan trials, which are based on the presenting cluster of target behaviors). Such a practice can be a source of frustration for treatment teams and requires careful planning, discipline, and diligence. The case report detailed is one example of how many of the above issues are manifested in clinical practice, and how one team dealt with them.

Case Report

The subject of this report is a 42 year-old ambulatory, African-American male with limited verbal skills who has lived at a regional intermediate care facility for the developmentally disabled since age nine. Current diagnoses include severe mental retardation, rapid cycling bipolar disorder, major motor seizure disorder, temporo-mandibular joint disease, and episodes of hypothermia of unknown etiology. His mental retardation (Stanford-Binet Intelligence Scale IQ < 30; Vineland Social Maturity Scale social age equivalent two years, nine months) is due to an unknown prenatal influence. Due to the mental retar-

dation, this individual has limited verbal skills. Significant severe behavioral issues dating prior to admission have at various times been labeled as psychotic disorder-NOS, schizoaffective disorder, and cyclical affective disorder. He has an abnormal electroencephalogram, however a recognizable seizure has not been observed in more than five years, with the last witnessed event occurring at age thirty.

Based upon clinical hypotheses, a variety of behavioral management strategies as well as corresponding psychopharmacologic interventions were previously initiated, with no sustained improvement noted in index behaviors. Previous psychopharmacologic interventions included trials of carbamazepine (1200 mg/day), amitriptyline (100 mg/day), clomipramine (150 mg/day), mesoridazine (up to 400 mg/day), thioridazine (up to 800 mg/day), haloperidol up to 12mg/day (discontinued due to dystonia, drooling, and pill rolling movements), diazepam (30 mg/day), and fluoxetine (discontinued at 40 mg/day due to increased agitation/irritability/hyperactivity). A previous trial of lithium in 1990 was discontinued due to a documented potential association with anorexia. All medication trials were deemed to be of sufficient dosage and duration to assess efficacy. Agents were discontinued due to either a lack of sustained efficacy or intolerable adverse effects.

More recently, frequent and predictable cycles in the documented pattern of behavior/sleep began to emerge following the initiation of mesoridazine and paroxetine in May 1997 (Figure One). He remained on this regimen in combination with divalproex until July 2000. While receiving this therapy, the patient had at least four complete behavioral cycles per year, ranging from 36 to 61 days in length. Rates of the index behaviors of concern fluctuated from a high of

350 intervals a month to less than twenty a month. Sleep also varied, with the greatest disturbance in sleep occurring concurrently with elevation in other target behaviors.

At the time of the psychiatric referral and ensuing change in therapeutic strategy, the primary presenting behavioral issues were worsening of sleep disturbance and increase in inappropriate social behaviors (aggression, property destruction, fecal smearing, stripping clothing, urination in inappropriate places). Waxing and waning of these behavioral symptoms were occurring in a frequent and predictable pattern. Due to the chronicity of the behavioral issues, complicated by an acute exacerbation, and the lack of a sustained improvement from previous therapeutic interventions, the treatment team referred the case to the consultant psychiatrist for re-evaluation.

Outcome Assessment Measurement

The behavioral data was collected in an ongoing manner by the trained staff in the home and work site, with support and guidance from the case psychologist. Sleep data was recorded using a standard sleep chart on which staff noted the presence or absence of sleep every thirty minutes between the hours of 10:00 p.m. and 7:00 a.m. The occurrence of aggression, property destruction, smearing feces, and stripping clothing (collectively referred to as inappropriate social behaviors) were documented by occurrence during each thirty-minute interval (i.e. multiple occurrences in an interval recorded as one incident). Data on inappropriate enuresis was reported on a per incident basis during each 24-hour period. Psychometric evaluations such as the Akathisia Rating Movement Scale (ARMS),⁷ Dyskinesia Identification Scale Condensed User Score (DISCUS),⁸ Aberrant Behavior Checklist (ABC),⁹ and Brief Psychiatric Rating Scale-Developmental

Disabilities (BPRS-DD)¹⁰ were used as psychometric and side effect assessments during the treatment regimen.

The ABC is a 67 item general purpose clinical rating scale designed to measure drug treatment effects on aberrant behaviors in persons with developmental disabilities. Each item is scored using a three point Likert scale (none, mild, severe), and the items comprise five independent subscales (Irritability, Stereotypy, Social Withdrawal, Hyperactivity, Excessive Speech). A trained research assistant rated the subject weekly. The ARMS is designed to detect akathisia in those with developmental disabilities and was completed by trained research personnel at predetermined intervals. The DISCUS was developed to assess treatment related and withdrawal emergent abnormal movements in populations with developmental disability. The DISCUS is based upon fifteen items (assessing movements in seven body regions) that are reliable, stable, and sensitive to withdrawal dyskinesias as well as tardive dyskinesia. Each body region assessment is based on a five-point scale ranging from zero (not present at all) to four (severe movements). A persistent total score of five or greater is usually indicative of tardive dyskinesia.¹¹ Trained research personnel completed this assessment at predetermined intervals. The BPRS-DD is an adapted version of the original BPRS, constructed for use in patient populations characterized by diminished cognitive functioning, and in particular those with developmental disabilities. It is a fourteen behavioral item caregiver assessment rated on a Likert scale of zero (not present) to six (present and extremely severe), with the rating given being an average of the presence and/or severity of that symptom for the last week's observations.

Intervention

The baseline medication regimen consisted of mesoridazine, paroxetine, and divalproex. Based upon the repetitive, cyclic nature of the index behaviors, a provisional diagnosis of rapid cycling bipolar disorder was made and a treatment plan designed. Mesoridazine 75 mg/day was decreased to 25 mg/day, and discontinued after one week. Paroxetine 20 mg/day was titrated to 10 mg/day for one week and discontinued. The atypical antipsychotic agent olanzapine was initiated at 5 mg/day for three days, and then increased to 10 mg/day. Olanzapine was decreased to 7.5 mg/day due to excessive sedation, which resolved at the lower dose. Lithium was initiated three weeks later, and increased to achieve therapeutic serum levels (0.5-1.2 mEq/l), with an eventual daily dose of 1200 mg/day. Index behavioral outcome data was collected and reported per month (Table 1) from baseline through medication introduction/titration and maintenance phases of therapy. This continuum of behavioral outcome data allowed for comparison of outcome measures pre- and post-treatment changes.

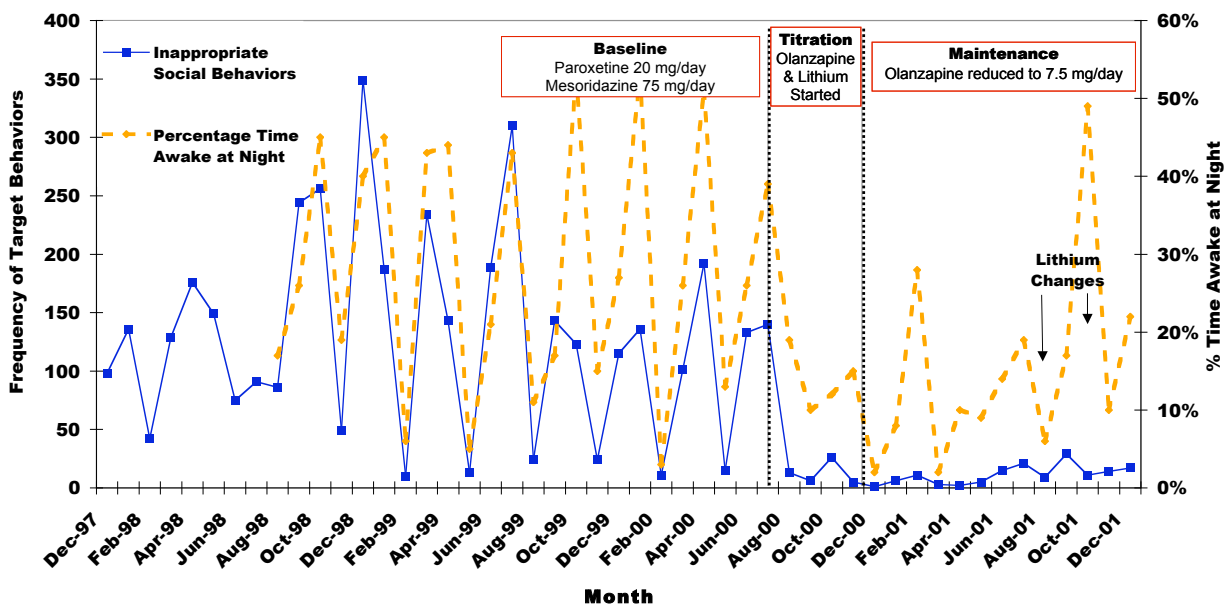
Results

Significant improvements in rates of index behaviors of concern as well as sleep patterns were noted after the initiation of the lithium, olanzapine, and divalproex medication regimen. Baseline labs prior to initiation of the lithium/olanzapine regimen included complete blood count, chemistry panel, urinalysis, and thyroid function studies and were unremarkable. Baseline calculated creatinine clearance was 91 mL/min. Maintenance phase trough valproic acid and lithium levels were 58.8 mcg/mL and 1.1 mEq/L, respectively. Lipids remained within normal limits throughout the period in review, as did blood glucose levels.

Baseline data collected prior to the medication changes indicated an average of 116.6 intervals of all measured index behaviors per month. Following the regimen changes beginning July 2000, data collected during the next five-month maintenance phase of treatment indicated a monthly average of 11.4 intervals for all measured target behaviors per month (Figure 1). Specifically, property destruction, aggression, stripping, smearing feces, incidents of enuresis, nighttime hours slept improved in a clinically relevant manner, compared to baseline measures (Table 1). The ABC scores improved from baseline to the final maintenance phase assessment (Table 2). ARMS and BPRS-DD scores did not change during the medication trial, while DISCUS improved from early maintenance to the last assessment (Table 3).

individual with severe mental retardation and an eventual diagnosis of rapid cycling bipolar disorder, as well as the difficulties encountered in arriving at this diagnosis. The initial diagnostic hypothesis was that of schizoaffective disorder, based on the presenting target behaviors. However, the response to the initial intervention was not satisfactory, leading to a re-examination of the diagnosis and corresponding treatment plan revisions. A review of the target behaviors showed intervals of waxing and waning sleep disorder and antisocial behaviors, with frequent periodicity. A thorough medication history revealed that the anecdotal reports of a previous adverse event with lithium therapy were not well documented, and a causal relationship between lithium (at therapeutic levels) and anorexia was not clearly established. This finding

Figure 1



Discussion

This case report reviews the effects of lithium, olanzapine, and divalproex combination pharmacotherapy on the behavioral indicators displayed by an

highlights the importance of thorough and accurate documentation of suspected adverse medication events, as well as the critical nature of a complete and accurate medication history when evaluating potential therapeutic options. It is the standard

treatment practice at this facility to change only one treatment parameter at a time, in keeping with the practice standards outlined by Rush and Frances², as well as Reiss and Aman.¹² However, the current case severity dictated the need to quickly and thoroughly re-evaluate the treatment regimen.

Table 1: Behavioral Outcome Data

Behavioral Parameter	Baseline: Mean (range)	Post-treatment change: Mean (range)
Property Destruction*	66.5 (11-191)	0.4 (0-3)
Aggression*	14.4 (0-50)	0.2 (0-2)
Stripping*	34.3 (0-69)	1.5 (0-13)
Smearing of feces*	7.4 (0-28)	0.4 (0-2)
Enuresis*	36 (11-76)	8.6 (1-23)
Sleep (night-time hours 2200-0700)	72%	85%

* by occurrence in 30 minute interval

As noted in Figure 1 and Table 1, significant improvements were seen in rates of index behaviors and in sleep disturbance, with the conversion to a more traditional bipolar disorder psychopharmacologic management plan. Additionally, the apparent period of stability between each behavioral cycle has increased/lengthened (no cycles documented in the five month maintenance period compared to at least four cycles per year in previous two years). The ABC data (Table 2) supports overall improvement; however the ARMS, DISCUS, and BPRS-DD data, while indicating stability during treatment, suffer due to lack of a baseline assessment for comparison (Table 3).

Table 2: Aberrant Behavior Checklist Assessments

ABC Scores / Phase	Irritability	Lethargy	Stereotypy	Hyperactivity	Speech
Baseline	2.2	1.1	1.4	2.1	2.8
Titration Phase	1.9	1.4	2.9	2.9	2.5
Maintenance Phase	0.1	0.06	0.8	0.06	0.8

Table 3: Other Psychometric Assessments

Psychometric Assessment Maintenance	Week*	DISCUS	ARMS	BPRS-DD
	12	7	1	9
	14	7	1	9
	17	8	0	22
	27	9	0	3
	31	0	1	6

*no baseline data obtained

It is quite possible that the discontinuation of paroxetine may be primarily responsible for this documented improvement, as SSRIs, like tricyclic antidepressants, may facilitate rapid cycling or hypomania.^{4,13} However, a number of critical treatment variables were altered at the same time period, making a clear association somewhat tenuous. The patient did have a previous adverse experience with the SSRI fluoxetine at 40 mg/day, in which discontinuation was required due to increasing irritability, agitation, and hyperactivity, giving increased credence to the hypothesis of an SSRI driven mood instability.

Of greater importance than the attenuation of a specific target behavior is to provide for holistic improvement in the patient's overall well being. In the present case, the patient's

observed quality of life and functional ability improved significantly with improved mood stability. The subject is now able to actively participate in his activities of daily living, work and earn pay, and spend it on preferred items rather than replacing property he had destroyed. He is more consistently able to participate in preferred community outings.

Conclusions

One cautionary note is to avoid inferring a psychiatric diagnosis based solely upon a response to medication. As each psychoactive medication may have multiple uses, this

strategy often leads to inaccurate diagnostic labeling. The importance of reviewing the current medication regimen as a potential contributor to the presenting problem is imperative, along with a corresponding detailed psychopharmacologic medication history. This case illustrates some of the many potential diagnostic and treatment issues encountered in treating psychiatric disorders in those with severe-profound developmental disability, and highlights the importance of clinical perseverance via a well organized, carefully planned treatment approach, using objective and measurable outcome parameters.

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