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Childhood-Onset Schizophrenia: The Challenge of Diagnosis

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Abstract

During the past two decades, the Child Psychiatry Branch at the National Institute of Mental Health has conducted a longitudinal study (including long-term prospective follow-up) of childhood-onset schizophrenia, a rare form of the disorder. Critical to this research has been accurate diagnosis. Outpatient screening has accurately diagnosed 55% of the 121 childhood-onset schizophrenia patients in the study to date. However, inpatient observation including drug-free observation has proven crucial to ruling out 96 children with alternative diagnoses who had been provisionally admitted for inpatient study. Standardized clinical ratings from outpatient screening only predicted 62% of these nonschizophrenia patients. Historically, medication-free observation was standard clinical care for difficult and unusual patients; this should be employed when possible in similar situations.

Keywords

Schizophrenia; Childhood-onset schizophrenia; Psychiatric diagnosis; Drug-free observation; Childhood bipolar disorder

Introduction

Childhood-onset schizophrenia (COS), characterized by onset before age 13 years, has a prevalence of approximately 1 in 40,000. This is a severe form of the illness with gradual onset and poor outcome. The psychotic symptoms that are the hallmark of schizophrenia are present in many alternative diagnoses (ADs).

During the past two decades, the Child Psychiatry Branch COS study has received more than 3,000 referrals, from which more than 350 children were screened in person. A total of 217 children were admitted as inpatients, 121 of whom have a finalized diagnosis of COS. We reported in 1994 on the status of the first 71 cases screened (COS $n=19$, AD $n=52$) [1]. During the following decade, we demonstrated continuity between child and adult versions of schizophrenia and uncovered the greater neurodevelopmental burden underlying this disorder [2]. To better understand the factors involved in validating a COS diagnosis, we provide an overview of the salient clinical features of 93 children admitted over the past decade.

Initial Screening

We selected children most likely to have COS through prior review of medical records followed by a full day outpatient screening by a team of two or more child psychiatrists, social workers, and nurses. Although all the children screened have many reports of psychoses in their medical/academic records and parental reports, we were able to rule out children with ADs during screening due to the sporadic nature of the hallucinations reported (often occurring only under stress), the lack of pervasive thought disorder, and a relatively intact social and cognitive presentation. It is clear as these children move through the medical system that psychotic symptoms, even when qualitatively and quantitatively mild, are highlighted and emphasized. This occurs despite the common knowledge that limited, transient psychotic symptoms are common across a broad spectrum of childhood disorders and in healthy controls; indeed, psychotic symptoms in AD disorders such as bipolar or major depressive disorder in children are found in up to 25% of these cohorts. We ruled out 38% of all children screened as having ADs with outpatient screening.

Medication-Free Period

Of the remaining 93 children admitted with provisional diagnoses of COS during the past decade, 30 (32%) were found during an inpatient observation that included a medication-free period to have an AD. These patients fell into four broad diagnostic categories: autism spectrum disorders ($n=9$), anxiety disorders ($n=7$), mood disorders ($n=7$), psychotic disorder not otherwise specified ($n=5$), and a single child best described as having childhood disintegrative disorder. It also should be noted that all the AD children suffer from one or more comorbid diagnoses in addition to their final primary diagnoses. The main reasons for our failure to detect AD children at screening appear to be related to nonspecificity of psychotic symptoms, the benefits of medication for these symptoms, and the diagnostic limitations of our clinical rating tools.

Because virtually all the children screened were on antipsychotic medication and approximately 20% also receive antidepressants, mood stabilizers, and/or anxiolytics, screening presentation was heavily influenced by medication. Atypical antipsychotics are widely known to be effective at treating and/or augmenting existing treatments for depression, bipolar disorder, anxiety, and obsessive-compulsive disorder, thus greatly adding to the importance of the drug-free observation. During the medication-free period, some children became manic without psychotic symptoms (bipolar diagnosis). Another child who walked head down in the halls and would not respond to peers/teachers/parents was found to have very severe complex counting obsessions, leading to psychotic-appearing compulsions (obsessive-compulsive disorder diagnosis). Yet another child whose poor social skills and odd fascination with Pokémon-type characters that were mistaken for negative symptoms and thought disorder was in fact judged to have Asperger's syndrome. Long-term follow-up has validated these exclusionary judgments in most cases.

We examined the clinical rating scales for psychosis, mood, and overall functioning collected at outpatient screening to see if they in fact help predict final diagnostic status at the medication-free period. We found that clinical rating measurements were only moderately useful for ruling out ADs. The best (though weak) rating scale predictors of diagnostic outcome for ADs are general severity ratings of psychosis (lower scores) and depression (higher scores). It is the severity, not the presence of psychosis—particularly auditory hallucinations, flattened affect, increased latency of speech, anhedonia, and social inattentiveness—seen in the COS children at screening that differentiates the COS children from the AD children. Conversely, the COS children report and rate significantly lower on levels of depressed mood compared with the AD children. When we analyzed 20 global

measures of clinical ratings that we routinely collect, it was the Bunny-Hamburg Psychoses and Depression ratings together that best predicted ADs (62% accuracy at screening, 85% accuracy at the medication-free period) [3].

Conclusions

Ruling out COS in severely ill, medicated children is particularly difficult for clinicians who see few COS children. An early and accurate diagnosis of COS will have profound implications for treatment. Conversely, ruling out a COS diagnosis has been equally important for future planning.

While clinical rating scales have utility in a research setting, they appear to have limited usefulness as a diagnostic tool when used to screen severely ill, medicated children with psychoses, and they are not a substitute for an astute clinician.

Our experience demonstrates that a medication washout is very informative; we recognize that it is increasingly difficult to accomplish this in clinical practice. When possible, however, putative, early-onset psychosis patients should be observed medication free, and one must be particularly circumspect about the diagnosis of COS in children with depression and/or sporadic hallucinations.

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