

# A Rare Condition in Child Psychiatry Practice: Childhood Disintegrative Disorder

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## ABSTRACT

A rare condition in child psychiatry practice: childhood disintegrative disorder

Childhood disintegrative disorder is a very rare pervasive developmental disorder characterized by normal development of verbal and nonverbal communication skills, social interaction, play, bladder and bowel control and motor behavior at least in the first two years, followed by regression between 2-10 years of age in two or more of the mentioned developmental areas. Estimated prevalence of the childhood disintegrative disorder is around 1.1-6.4 per 100.000 children and the common age of onset is 3.36 years. The present report describes a boy who developed childhood disintegrative disorder after approximately 3 years of normal development and clinical features are discussed in the light of the clinical literature.

**Key words:** Childhood, disintegrative disorder, pervasive developmental disorder

## ÖZET

Çocuk psikiyatrisi pratiğinde nadir bir durum: Çocukluk çağı dezintegratif bozukluğu

Çocukluk çağı dezintegratif bozukluğu, yaşamın ilk iki yılında sözel ve sözel olmayan iletişim, sosyal etkileşim becerileri, oyun, mesane ve barsak kontrolü ile motor davranış alanlarında normal gelişimi takiben 2-10 yaşları arasında belirtilen alanların en az ikisinde gerileme izlenen ve son derece nadir görülen bir yaygın gelişimsel bozukluktur. Tahmini prevalansı 100.000'de 1.1 ve 6.4 arasında değişmektedir ve ortalama başlangıç yaşının 3.36 olduğu ifade edilmektedir. Bu yazıda üç yıllık normal gelişimin ardından çocukluk çağı dezintegratif bozukluğu belirtileriyle başlayan bir erkek olgu sunulmuş, klinik izlemi ve klinisyenlere verebileceği ipuçları literatürdeki bilgiler ışığında tartışılmıştır.

**Anahtar kelimeler:** Çocukluk çağı, dezintegratif bozukluk, yaygın gelişimsel bozukluk



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## INTRODUCTION

Childhood disintegrative disorder (CDD) is defined as a neuropsychiatric syndrome in which verbal and non-verbal communication, social interaction skills, play, fecal and urinary control and motor behavior are normal in the first two years of life but followed by regression at least in two of these fields between 2-10 years. Clinical manifestations may be accompanied by qualitative impairment in social interaction and communication, a restrictive, repetitive and stereotyped pattern in behaviors, interests and activities (1). While CDD takes place under the heading of "Other Childhood Disintegrative Disorders" in order to discriminate from Rett Syndrome in ICD-10 (2), it is classified under the heading of "Pervasive Developmental Disorders" in DSM-IV (3). In literature, CDD is also named as disintegrative disorder, Heller's dementia or Heller's

syndrome. Some changes were done for diagnosis category in the recently published DSM-V. In DSM-V, use of "Autism Spectrum Disorder" was proposed as a unified single diagnosis instead of autistic disorder, Asperger's syndrome, pervasive developmental disorder not otherwise specified and CDD which are defined under the heading of "Pervasive Developmental Disorders". Rett's syndrome was not included in the diagnosis due to its genetic background (4).

CDD is a quite rare disorder of which etiology is not clearly known yet (5). Its prevalence was shown to vary between 1.1 and 6.4/100 000 (6,7). CDD is reported to be more prevalent among boys than girls although exact ratios were not detected (7,8). Mean age of onset is 3.36 years. However cases aged between 1.2 and 9 were reported in literature (8).

CDD may begin insidiously (within months) or acutely (within days) (9). "Premonitory period" which

includes precursor symptoms like anxiety, hyperactivity and irritability may be detected before onset of regression in functionality in some part of the cases (9). The disease may be confused with many clinical conditions like anxiety disorders, depressive disorder, attention deficit and hyperactivity disorder during this “premonitory period”.

In this paper, a boy aged 3 years and 3 months who was being followed up at our clinic is presented due to CDD which is a rare condition.

## CASE

The 39 months old boy was admitted to our outpatient clinic with complaints of “inability to speak, hyperactivity and restlessness”. It was learned from his mother that, he was hyperactive, he could not stand in his place even one moment and continuously turned around himself, he was very irritable and nervous, he did not reply when his name was called, his eye contact was limited, he did not speak, he communicated with nobody and he did not play with his peers and toys.

In his medical history, he was born from the second pregnancy of the mother, he was delivered with normal vaginal delivery on term, birth weight was 3150gr and there were no problems during pregnancy, delivery and after birth. It was stated that he had a healthy sibling three years older than him, and there was no loss of fetus, curettage or early baby-child deaths in the family. When neuromotor development of the child was questioned, he was learned to be fed with breast milk until one year, walk at one year, begin to make sentences at two years, to inform about his micturition and defecation during the day since 2.5 years and to wear napkin at night, play with his peers and toys and not to have a communication problem. His first complaints began three months ago although he had no developmental problems until 3 years, he was learned to wake up suddenly one night by crying and screaming, he had fears that his family could not understand since that night and his sleep patterns impaired. Then it was learned that he had hyperactivity, he continuously turned around himself, he was not able to talk, he did not inform about his micturition and defecation, he

was nervous, he did not communicate, he did not reply when his name was called and a progressive regression occurred in his development.

His family history was unproblematic. The mother was 29 years old, graduate of lycee, housewife; the father was 31 years old, graduate of high school and he was a civil servant. It was learned that there was not consanguinity between parents, there were no family members with any psychiatric, neurologic or metabolic diseases and developmental disorder. The patient relation with his 6 year-old healthy brother was reported to be good until the onset of the disease however it impaired thereafter. Attitudes of the parents were seen to be normal.

It was learned that the patient had been initially admitted to a pediatric neurology clinic, he was evaluated for metabolic diseases, his blood tests were normal and thyroid function tests were normal. No pathologic findings were found in sleep awakenings electroencephalography (EEG) and cerebral magnetic resonance imaging (MRI) and his neurologic examination was also found normal. He had been referred to our clinic thereafter.

On psychiatric interview, he was observed to be conscious however he did not have eye contact, he did not reply when his name was called, he did not obey the orders, he did not speak, he moved during the interview and he continuously turned around himself in the middle of the room.

The patient was considered to have CDD according to DSM-IV criteria as his development looked normal within the postpartum two years, he lost his abilities like verbal expression, social skills, fecal and urinary control, play, motor skills before ten years; restrictive, repetitive and stereotypic patterns of behavior including motor stereotyped behaviors and manierism, and this disorder could not be better explained with other pervasive developmental disorders (PDD) or schizophrenia. It was considered that the patient could be evaluated as CDD through being in contact with pediatrics clinic. Risperidone 0.5mg daily was started. The family was given psycho-education and the patient was recommended individual and group education and invited for control three weeks later. He was recommended to go to pediatrics follow ups

concurrently. The patient did not come to the next control. He was admitted about four weeks later without appointment. On that admission, they stated that they could not begin special education due to ongoing bureaucratic procedures, therefore they could not come for control however they used the drug regularly and benefited partially. The patient was detected not to have any problems on organic examinations however his eye contact was seen to partially increase and better obey the orders. No side effect of the drug was observed. Behavioral advices and more detailed information about the nature of the disease and responsibilities of the parents were given to the family and risperidone dose was elevated to 0.75 mg daily. On his next control, it was stated that he began special education, experienced mild adaptation problems at school however they resolved thereafter and his eye contact was seen to increase. He is being followed up by us.

## DISCUSSION

In this paper, a male patient aged 39 months who was followed up with diagnosis of CDD is presented. CDD is a disorder which is very rare in pediatrics practice and of which etiology is not fully known yet (5). Literature about CDD is mostly composed of case reports (9-13). CDD may begin at approximately 3 years incidentally or acutely (within days) (8,9). A "premonitory period" which includes precursor symptoms like anxiety, hyperactivity and irritability may be detected before onset of regression in functionality in some part of the cases (8). In our case, the disorder has begun acutely around 3 years and the family was admitted to neurology clinic by considering an organic disorder. It was concluded that the patient did not have an organic pathology as the result of the tests so the patient was referred to our clinic and evaluated as CDD according to DSM-IV criteria and exclusion method. Complaints of anxiety, sleep disorders and hyperactivity was seen to also indicate the features of "premonitory period" mentioned in the books. This condition made us to consider that "the child noticed the regression in himself however he could not express this so he reflected this condition to his behaviors". Afterwards, reduction in

awareness ability together with the loss in other abilities may be reducing the findings of "premonitory period".

In literature, while speech is observed to be the mostly impaired skill, it is followed by social skills and adaptation behaviors, stereotyped behaviors, loss of fecal and urinary control and loss of motor skills (9,14,15). In our case, similar losses were detected and the patient was immediately directed to special individual and group education without stopping follow up in pediatrics clinic. The patient experienced adaptation problems in the first days of special education however these problems rapidly decreased and disappeared thereafter and he was seen to respond to special education together with medication. Although the loss in mental skills is reported to be permanent in literature (16), mild improvement in our case suggested that early thinking of diagnosis of CDD, early starting supportive intervention although "all organic examinations have not been completed yet" could reduce the rate of skill loss.

Before making the diagnosis of CDD, a detailed neurologic analysis should be done in order to discriminate neurologic disorders, chronic neuro-infections or epileptic encephalopathies and seizures that may accompany to CDD (15). In our case, no pathologic findings were detected in neurologic examination, routine blood tests, thyroid function tests, cranial MRI and EEG done in pediatric neurology clinic and organic etiology was not considered. Other pervasive developmental disorders should be considered in differential diagnosis, particularly autism should certainly be discriminated from CDD. CDD may be discriminated from autism by neuromotor developments in the first 2 years of life being normal and absence of a progressive skill loss. Symptoms begin before 3 years in autism. Speech development is seen not to be sufficient when symptoms begin. Motor skill impairment is usually not seen. Acquired loss of anal and urinary control is not seen and a progressive destruction is not seen in cognitive functions (16). Therefore autism was not considered in our case.

CDD does not have a specific treatment. Psychoeducation of the patient and the family, special education and psychopharmacologic agents are the

main treatment methods. Psychopharmacologic treatment option is frequently considered particularly in presence of anxiety findings, hyperactivity, self-destruction, aggression, behavioral problems. Atypical antipsychotics (11,17-20) and selective serotonin reuptake inhibitors (20) are seen to be used frequently for this purpose. In our case, risperidone treatment was started for nervousness, behavioral problems and hyperactivity complaints, consistently with literature, and also the patient was directed to special education in order to achieve basic skills after the family had been given appropriate psychoeducation. On the last control, the patient was seen to partially benefited from

treatments. This condition emphasized the importance of early intervention for diagnosis and supportive therapy once more.

CDD is the least known pervasive developmental disorder. Diagnosis of CDD may be overlooked due to absence of a specific diagnostic test, the symptoms resembling autism and initial findings may be confused with many other psychiatric disorders. Therefore reporting more clinical cases and larger studies with longer duration and follow up studies would help us to understand the emergence type of the disorder and to early detect CDD and make interventions in the early period.

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