



# Children with neurodegenerative disease and obsessive-compulsive behaviour

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KRISTIN ROMVIG ØVERGAARD

E-mail: kristin.romvig.overgaard@ous-hf.no

Division of Mental Health and Addiction, Oslo University Hospital  
and

Department for Paediatric and Adolescent Medicine, Akershus University Hospital

Kristin Romvig Øvergaard, PhD, child and adolescent psychiatrist and researcher.

The author has completed the ICMJE form and declares no conflicts of interest.

BEATE ØRBECK

Division of Mental Health and Addiction, Oslo University Hospital

Beate Ørbeck, PhD, clinical psychologist and researcher.

The author has completed the ICMJE form and declares no conflicts of interest.

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## BACKGROUND

Children with neurodegenerative diseases progressively lose skills and develop somatic and psychiatric symptoms. Obsessive-compulsive disorder (OCD) may occur, a disorder for which effective medical and psychological treatments are available. However, whether these treatments are useful for children with neurodegenerative disease is unknown.

## CASE PRESENTATION

A child with an uncommon form of neurodegenerative disease (with loss of sight and incipient cognitive decline) had during the previous year developed time-consuming compulsive behaviours. The child spent much of the day at home (> 8 hours), turning on/off switches, opening/closing doors, repeating after others and so on. A diagnostic assessment concluded that the child fulfilled the criteria for OCD, with predominantly compulsive acts. The child began combined treatment with sertraline and exposure and response prevention therapy (ERP). It was necessary to adapt the ERP to the neurodegenerative disease, with strong parental involvement. After six months the compulsive acts were gone.

## INTERPRETATION

Children with neurodegenerative disease may have a high risk of psychiatric disorders, but the literature is sparse on phenomenology and treatments. The present case study documents that OCD can occur in a child with neurodegenerative disease and that the combined medical and psychological treatments were effective.

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Children with neurodegenerative diseases progressively lose skills and develop new somatic and psychiatric symptoms. Recurring obsessive thoughts and compulsive acts may occur. There is effective treatment for this disorder, but it is not known whether this kind of treatment benefits children with neurodegenerative disease.

A child was brought by its parents for a consultation at the local hospital after they had observed an increasing need for control over the past year. The child insisted on performing many acts itself, in very definite ways and repeatedly. The acts were particularly connected to light switches, mealtimes, dressing and mimicking others. All three were worn out from spending most of the day on these acts. When the parents asked the child why it insisted like this, the answer was: "It's my body that decides on it". Three years previously, an assessment had confirmed a rare neurodegenerative disease.

There had been a gradual reduction over a period of years in the child's vision (now reduced to lateral vision), motor function and cognition (particularly language, memory and concentration).

Assessing symptoms of psychiatric disorders in children with neurodegenerative diseases is challenging, as the primary disease itself is a strain on the patients and their families. Most of the patient's feelings (for example sadness, anxiety, frustration, anger and loneliness) can be understood as natural reactions to limitations due to loss of function and ever new symptoms. Clinicians may doubt whether it is meaningful to describe the symptoms as one does for otherwise healthy children. In addition, the patients often experience such burdens as loss of friends, changes in their everyday school life, dependence on aids and regular contact with the health service. Taken as a whole, neurodegenerative diseases are not uncommon, but each of the individual diseases is. Clinicians will therefore often have too little experience of them. Limited knowledge and limited available literature on psychiatric disorders associated with neurodegenerative diseases makes assessment demanding and the diagnosis uncertain.

The parents explained that the problems had begun gradually, and that it took time before they thought of the acts as anything other than the child wanting to be independent. For almost a year before they came to the first consultation, the acts had taken up a great deal of time, and in the last couple of months they had lasted through most of the waking day. In the morning the child insisted on turning on the light a particular number of times. If the parents did it, everything became difficult, and the child then had to turn the light on and off for a long time. Similarly, there were specific acts associated with getting dressed and mealtimes so that if the parents for example put out the child's shoes or put food on the plate, the whole dressing routine or meal had to start again.

The child also mimicked much of what its parents said and did. This mimicking became demanding because of the child's impaired vision and cognitive disabilities. If the child did not catch everything, it became distraught and insisted that the parents repeated it. The echopraxia led to the family being unable to carry on an ordinary conversation. The child was also particularly obsessed with turning on and off switches, going in and out of doors, opening and closing doors, going to the toilet repeatedly, and insisting on exact positioning of objects and precise performance of daily routines. The child threw itself down, used bad language and hit out in all directions, distraught, when these needs were not met. The parents felt that they really needed to just sit quietly at home to avoid making mistakes in relation to these demands. They often dropped the child off at school late and got to work late themselves. In the evening they spent hours getting through bedtime routines.

At school the child showed a tendency to repeat after its regular assistant, but that did not disturb its classwork. There was also little obsessive behaviour outside the home in other respects. Neither the patient nor the parents could explain the reasons for this, but perhaps the acts emerged in the most secure surroundings, and when there was most time to repeat acts. The child sometimes talked about its own concern that something might happen to its parents, but denied that such thoughts were recurring. As part of the assessment, the patient, with help from its parents, answered the questionnaire Children's Obsessive

Compulsive Inventory (CHOCl) (1). Only compulsive acts emerged here, consuming an estimated > 8 hours daily. There were no other symptoms of psychiatric disorders, such as anxiety, depression or behavioural problems.

The description of the symptoms raised suspicion that the patient fulfilled the criteria for the ICD diagnosis Obsessive-compulsive disorder (OCD): F42.1: Predominantly compulsive acts (2). The patient was unable to mention repetitive obsessive thoughts, but was able to describe a feeling/thought of having to perform them. Although the descriptions and volume left no doubt that the criteria were met, there was still some uncertainty as to whether the acts were linked to underlying anxiety, as they most frequently are in somatically healthy children.

The compulsive acts also occurred to only a limited extent outside the home. Some of the acts were moreover perceived as of benefit, i.e. as a desire to be independent despite loss of function. Impaired vision and cognitive disabilities also demanded special preparations with fixed routines and particular location of objects to make everyday life manageable. As a result, the parents gradually participated in more and more behavioural patterns. In the same way as parents with otherwise healthy children with OCD, these parents, too, participated in the obsessive acts (accommodation) in order to avoid upsets resulting in further delays in daily routines. Nonetheless, the patient's neurodegenerative disease and the fact that the parents would be responsible for conducting all exposure exercises gave rise to doubt as to whether treatment would be of benefit.

A combination of medical management and gradual exposure with response prevention (ERP) was started (3). The patient was put on 25 mg sertraline (Zoloft) daily for a week. This was then increased to 50 mg, which became the maintenance dose. Both patient and parents received psychological therapy with focal areas described in Box 1. There were no therapist-assisted exposures, only guidance for the parents, who conducted the exposures.

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## Box 1 Focal areas for the psychological treatment

### 1 PSYCHO-EDUCATION

The patient and the parents received information about OCD, including accommodation (that the parents go along with the obsessive acts)

### 2 GRADATION

The symptoms were reviewed in detail. Together, the patient and the parents graded the acts according to how difficult they were to resist (used 'steps' as an illustration of the degree of difficulty). Within each general area (e.g. turn light switches on and off), the degree of difficulty was assessed further (different light switches in different rooms). They were given the task of starting exposure by practising what they thought was simplest (stop turning on and off the switch the patient was least obsessed with), to ensure success and motivation for further exposure

### 3 AWARENESS-RAISING

To make the patient aware of how difficult it was to resist acts, and experience that it could become less difficult in the course of the exposure exercise, the parents were supposed to help to grade the distress by visualising a thermometer (graded from 1-10) during the exercises

### 4 TOOLS

Patient and parents discussed which tools could be used for the child to manage to withstand the distress that arises when acts are not carried out (use of iPad, talk about favourite animals etc.)

## 5 EXPOSURE WITH RESPONSE PREVENTION AND HABITUATION

The patient was supposed to do the exposure exercises long enough for the distress of not performing the acts to actually subside, i.e. the act they were working on at a particular time didn't 'have to be done'

## 6 REWARDS

The child and the parents agreed on short- and long-term rewards

## 7 CLOSE PARENTAL MONITORING

In addition to the daily work, the patient and parents had short weekly meetings at which they reviewed progress and challenges and agreed on rewards. Prior to the meetings, the parents had agreed between themselves what they would talk about, because the patient could not manage lengthy conversations about the topics. The parents could contact the therapist by telephone. The therapist called the mother weekly during the initial period after the start of the treatment. The first follow-up at the hospital was after one month

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Children with OCD are usually first given psychological therapy in the form of ERP, then later, if this is not effective, medical management (3). In this case drug treatment was started immediately, and sertraline, a selective serotonin reuptake inhibitor, was chosen. Sertraline is assumed to have an effect on obsessive thoughts and acts by specifically inhibiting serotonin uptake in the neuron terminals and is indicated for OCD in children and adolescents. At the same time, psychological therapy was administered.

There were several reasons for choosing combination treatment. Because of the neurodegenerative disease, the patient was unable to participate in the treatment in the same way as a somatically healthy child (for example weekly appointments) and it was initially uncertain whether it was possible for the child to focus on the exposure tasks. The impaired vision made it impossible to use visual aids, which as a rule are useful in ERP for children, particularly those with cognitive disabilities. It was uncertain how soon there would be loss of function and/or new symptoms. In addition, the volume of the obsessive acts consumed all the time at home in a family that already had many care tasks. The hospital also had limited capacity for close follow-up. The treatment was therefore dependent on closer parental control than was usual, even though the parents had regular telephone contact with the therapist. Emphasis was placed on the parents regularly having short meetings with their child at which they made concrete agreements on steps in the exposure and how they should help in the exercises. They had to use verbal reminders and visualise images in their communication. Despite the adaptations made to the treatment, it was still uncertain whether it would be of benefit.

In order to estimate the efficacy of the therapy during the process, the patient and the parents answered a question on the time spent on obsessive acts. Prior to the treatment, the child spent > 8 hours a day on the acts. Two–three weeks after the start of treatment, one of the obsessive acts stopped spontaneously (toilet visits), possibly as an effect of sertraline. One month into the treatment, the compulsive acts were substantially reduced (to 3–4 hours a day). The patient had then been exposed to resisting turning light switches on and off. In the beginning, the parents thought it was difficult to spend so much time on one switch, but when they saw the mastery achieved, it became meaningful. They had short weekly meetings at which they verbally agreed on the plan for the week, the tools to be used and rewards for completion. One reward the child chose was to eat at a restaurant. After six months, less than one hour a day was spent on obsessive acts. See Table 1 for scores before and after therapy for the CHOCI questions where the patient had a high score before therapy.

**Table 1**

The patient's scores on Children's Obsessive Compulsive Inventory (CHOCI), before and after treatment (only statements where the patient had a score for symptoms). Scale: 1 = not, 2 = a little, 3 = a lot

CHOCI statements	Before	After
I feel that I must do ordinary everyday things in exactly the same way each time I do them	2	2
I check things like taps and switches over and over again	3	1
I get very upset if my things are not always in exactly the same place	2	2
I often get behind in my school work because I write the same words over and over again	2	1
I spend a lot of time every day checking things over and over again	3	2
I often have problems getting things finished because I need to be absolutely sure that everything is exactly right	3	2
I check over and over again that my doors or windows are shut, even though I try not to do it	3	1

The family found that by spending time on the simplest tasks at the beginning, it became easier to resist the most difficult compulsive acts. They did not think there was anything left to practise after six months, as the acts were no longer being repeated (Box 2). They could see nonetheless that the child could become obsessed with various click sounds, and that transitional situations required a lot of time. They noticed a tendency to repetition of acts when the child was bored, which caused some concern about a possible relapse. However, the parents felt secure in that they now had tools available if the obsessive acts should return, including continuing the short weekly meetings. The child said itself that it was particularly useful to use 'steps', rewards, weekly meetings and thermometer (Box 1)

### Box 2 The patient's advice to other children with OCD

"When you feel a thought that you sort of have to do it. Get used to it being there, and try and tackle it. Then you'll fix it!"

## Discussion

Neurodegenerative diseases cause progressive loss of neurons in the central nervous system and may result in gradual loss of vision, motor function and cognition (language, memory, concentration) after a period of normal development (4). Epilepsy is also common during the course of the disease, but this patient had not so far had suspected seizures. The cumulative incidence of neurodegenerative diseases in children is calculated to be 0.6 per 1 000 live births (5).

In order to fulfil the criteria for OCD, a patient must experience repeated, unwanted and disagreeable thoughts and/or repeated, non-useful acts or rituals that are performed to alleviate anxiety, often to prevent specific events, such as an accident or injury. If the compulsive acts are resisted, the anxiety is exacerbated (2). It is known that psychiatric disorders occur 3–4 times as often in children with cognitive disabilities as in children without these disabilities (6). Symptoms of various psychiatric disorders are reported to be common through the course of neurodegenerative diseases, but the literature consists to a large degree of case studies. Nevertheless, in a study of children with a subgroup of neurodegenerative diseases (n = 258, age 0–14 years), a high prevalence of behavioural problems was reported, but also of anxiety symptoms in 36–73 % of the children (varying for

different diseases) (7). A review article about another subgroup of neurodegenerative diseases reported psychiatric symptoms in the majority of the children and anxiety symptoms in 45% (8).

It has been pointed out that there is a need to develop treatment for children at high risk of psychiatric disorders, for example those with syndromes (9), but so far there is no concrete advice for clinicians with patients with specific rare diseases. We have to draw on studies of somatically healthy children or children with developmental disorders, for example. It may be useful for clinicians to know that a Norwegian practical manual has been developed that amongst other things recommends adaptations to psychological therapy for OCD in cases of developmental disorders (10). It is known that combined therapy with sertraline and ERP is an effective therapy for OCD in children (11), but whether it is more effective with sertraline in addition to ERP is unclear because there are few studies (12). It may thus be most effective to provide psychological therapy alone for a period, also to children with intellectual disabilities (13). If, for various reasons, medical management is nonetheless decided upon, there will still as a rule remain compulsive symptoms for which psychological treatment may be useful (11).

Offering ERP alone to children with cognitive disabilities implies both practical and theoretical problems. Treatment of psychiatric disorders in this group of children may receive inadequate priority in the healthcare service, with the consequence that children may be unnecessarily medicated. In addition, paediatricians who treat neurodegenerative diseases may lack expertise in psychological therapy. If they nonetheless provide such treatment themselves, guidance by a therapist with experience of OCD may be useful, not least if the treatment is not being effective. Clinicians often assume that children with cognitive disabilities will have difficulty in following a therapy. This despite the fact that a review article concluded that at least those with a milder degree of disability may benefit from psychological therapy (14).

Consistent with this study, earlier case studies have also shown that it is possible to treat children with cognitive disabilities, including OCD (15). To achieve successful treatment of children with additional disabilities, it is nonetheless important to adapt the treatment to the child's premises (9). For the patient in question, this meant less frequent consultations than usual, and that the therapy was conducted by the parents with guidance by telephone.

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