Rev Med Int Sindr Down. 2014;**18(3)**:43-46

INTERNATIONAL MEDICAL REVIEW ON DOWN'S SYNDROME

www.elsevier.es/sd

FUNDACIÓ CATALANA SÍNDROME DE DOWN
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CLINICAL CASE

Regression in young adults with Down's syndrome. A three cases review

B. Garvía[a,*] and B. Benejam[b,*]

[a]*Specialist Psychologist in Clinical Psychology and Psychotherapy, Centro Médico Down, Fundació Catalana Síndrome de Down, Barcelona, Spain*
[b]*Specialist Psychologist in Neuropsychology, Centro Médico Down, Fundació Catalana Síndrome de Down, Barcelona, Spain*

Received 5 May 2014; accepted 6 June 2014

KEY WORDS
Down's syndrome;
Depression;
Regression;
Loss of skills

Abstract
A deterioration in some of their cognitive functions and adaptive skills has been observed in adolescents and young adults with Down's syndrome (DS), which is similar to that observed in individuals who suffer from Alzheimer's disease, although at much earlier ages. Little is known of the etiology of these changes. In this work, 3 cases are presented on 3 young adults with DS, who presented with a deterioration in their cognitive functions and a loss of, previously well attained, functional skills. Some medical and psychological risk factors that could be associated with this regression are examined. In all the cases, the loss of skills was associated with anxiety and depression symptoms.

PALABRAS CLAVE
Síndrome de Down;
Depresión;
Regresión;
Pérdida de capacidades

Regresión en adultos jóvenes con síndrome de Down. Revisión de tres casos

Resumen
En algunos adolescentes y jóvenes con síndrome de Down (SD) se ha observado un deterioro en sus funciones cognitivas y en sus capacidades adaptativas similar al observado en personas que padecen la enfermedad de Alzheimer, aunque en edades muy tempranas. Se sabe muy poco sobre la etiología de estos cambios. En este trabajo se exponen tres casos de jóvenes con SD que presentaron un deterioro en sus funciones cognitivas y una pérdida de habilidades funcionales previamente bien adquiridas, y se examinan algunos de los factores de riesgo (médicos y psicológicos) que pueden estar asociados a dicho deterioro. En todos los casos, la pérdida de capacidades se acompañaba de síntomas de ansiedad y depresión.

*Authors for correspondence.
E-mail: beatriz@fcsd.org (B. Garvía);
bbenejam@fcsd.org (B. Benejam).

Introduction

Several authors have described severe regressions in adaptive and cognitive functioning in adolescents and young adults with Down's syndrome (DS)[1,2]. It has to be asked whether these regressions can be related to dementia of unusually early onset or due to depressive aspects. In 2011, the *International Review of Research on Developmental Disabilities* journal published an article by Devenny and Matthews titled: "Regression: atypical loss of attained functioning in children and adolescents with Down's syndrome". The text dealt with severe regressions that had been observed in some adolescents and young adults with DS as regards their cognitive and adaptive functioning, and which had appeared after a period of normal progression. The signs and symptoms consist of, on the one hand, an atypical loss of previously attained skills, in cognition, in socialization, and in daily activities and, on the other hand, an increase in disadaptive behavior. All the cases showed a behavior that appears within the description of depression: loss of happiness, isolation or withdrawal and crying, sadness and tiredness. The authors classified the appearance of this problem in young individuals as "rare", and they stated that very little is known about the problem, and that it is not clear if these cases should constitute a special diagnosis or form part of the psychopathology or of disadaptive behaviors[1].

DS is a risk factor for developing depression[3]. In emotional stress situations, and taking into account their current lower ability of understanding, perceiving and expressing their thoughts and emotions, individuals with DS may present with atypical, and more complex, responses compared to individuals with no intellectual disability[4]. Depression in this group is not usually expressed in words, but in the form of loss of abilities and memory, slowing down, inattention, changes in appetite or sleep rhythm, a deterioration in cognitive skills, a tendency to disconnect, and isolation, soliloquies and delirious ideas, affective lability, passivity or crying[5]. It is estimated that neuropsychiatric symptoms occur in approximately 26% of individuals with DS[6].

On the other hand, there is a close relationship between DS and Alzheimer's disease (AD) as a result of the overexpression of the amyloid precursor protein (APP) gene, present in chromosome 21 that, in DS, is triplicated[7]. Alzheimer's disease (AD) is probably the most important challenge that individuals with DS have to face. Several authors have described behavioral and personality changes as the first symptoms of AD in individuals with DS, among which are highlighted; apathy/lack of motivation, aggressiveness, emotional lability or social tact, depression, and the presence of delirious ideas[8-10]. Given that much of these symptoms, together with the sleep disorders and loss of appetite, are common to both depression and AD, it is important to make a good differential diagnosis between the two.

A prior baseline of anxiety, depression or aggressive behaviors have also been detected in the young adults that have presented with severe regressions, and the authors of the referenced article[1] pointed out four possible risk factors that could lead to regression: menarche, transition phases, life events, and exposure to anesthesia. Prasher[11], in 2002, observed that this regression lasted about two years, and it was followed by a stationary phase. He noted that some improvement could be observed with antidepressants, although the symptoms could persist. This author attributed the regression to three possible causes: premature changes associated with early onset of AD; disruption of routine, and loss of proper support in the transition from child to adult, and rupture of their identity on being aware of DS.

In the Centro Médico Down of the Fundació Catalana Síndrome de Down, with more than 2500 clinical records of patients with DS, we have dealt with a reduced number of these cases (and some others that have consulted us by e-mail, and providing documentation) that would have entered this clinic and that they coincided with the adolescent/young adult phase, with changes or transitions or becoming aware of the disability. We emphasize the rupture of their identity on being aware of their DS as a cause of regression.

Case presentations

Clinical case 1

A 22 year-old female who consulted after starting with a clinical picture of deterioration of one year onset, in the form of mutism, motor slowing down, social withdrawal, loss of interest, isolation, difficulties in orienting herself in time, and major problems in concentration. The parents also mentioned the presence of "odd habits" (not wanting to be touched, wetting her hair, hiding knives, etc.). As medical background of interest, is highlighted hypothyroidism on pharmacological treatment. After a detailed interview with the parents, it was observed, at the same times mentioned, that an important change in the life of the patient had occurred (a transition). She went from an ordinary school to an occupational workshop. It appeared to the family that this is when she started to get worse.

Previously, the patient expressed herself clearly and was able to explain without difficulty the activities she performed during the day. Later, she started to stutter, and months later stopped speaking (during this time she communicated by signs).

As regards daily skills, a slowing down was observed at first, and then she started to refuse to do things. She had stopped doing things that she liked, such as read, dance, and practice sport. At home she passed the time watching television or locked in the bathroom in the dark. To this symptomatology should be added a significant weight and hair loss.

The patient was referred to the general practitioner who, after an exhaustive examination, ruled out a medical problem. She was then referred to Neuropsychology, which ruled out the presence of a degenerative type syndrome on follow up. From the neuropsychology examination, a marked attention deficit was noted, as well as severe slowing down of psychomotor function. The cognitive profile of the patient supported the diagnosis of a depressive disorder.

The psychiatric and psychological examination also pointed towards a major depressive disorder, with clinical signs of hypothymia, dysphoria, isolation, psychomotor

inhibition, weight loss, apathy, anhedonia, mutism, signs of obsessive disorder, and severe regression.

After the administration of duloxetine 30 mg she began to improve, with a greater reactivity to stimuli, disappearance of psychomotor inhibition, disappearance of mutism, improved sociability, weight increase, and recovery of motivation. The compulsive symptoms persisted, as well as the inappropriate reactions (outbursts of anger) to frustrations, although she showed to be more content, increased her appetite, cared more for her appearance, and was more sociable.

This would be a case of regression due to depression that was triggered at a time of transition: change from normal school to special education, a critical phase, adolescence, and we believe that it has to do with perception/or assuming of identity and the disability.

Clinical case 2

A 26 year-old female, who consulted for the first time when she was 13 years-old due to having a rebellious and oppositional attitude. She was communicative, pleasant and likeable. She studied until the 6th year in an ordinary school and afterwards in an institute, sharing ordinary education with special education (two days per week). The entrance to the special school confronted her with the disability: "*I don't like them in this school because they are bad*", demonstrating a significant rejection of the syndrome. After a period of opposition and anger, she began a pseudo-delirious discourse. The parents explained that their daughter does not want to have DS. Sometimes she says that she wants to die, provoking and looking for confrontation.

When she was 19 years-old her brothers become independent and left home. This situation was a determining factor for her: she missed them, could not sleep, and said she wanted to be independent as well. She presented with depressive symptoms —sadness and crying— not looking into eyes, was afraid and insisted that she does not want to have DS. She is very concerned and irritable and talks a lot about her disability: "*where did I get this face from?*". She says that she does not belong to her family. She also forgets many things, is disoriented and is anxious. She started to have mutism, conciliation insomnia and a generalized slowing down.

The psychiatrist prescribed an antidepressant. The case was referred to a neuropsychology where a marked psychomotor slowing down was observed, along with lack of initiative/spontaneity and significant difficulties in understanding, secondary to an attention deficit. The clinical picture of the patient did not suggest a primary degenerative disease.

Far from an improvement with the medication and psychotherapy support, she is more "disconnected" and began to perform stereotypes, stutters a lot and her language is almost intelligible, until she began to stop speaking. She refuses to go to the occupational center, has lost skills (since is unable to dress herself alone). She was prescribed Risperdal® 1/2-0-1/2 and, although the stereotypes disappeared, apathy and disinterest persists. Given the great increase in weight with the medication, it was changed to Abilify® 5 mg 1-0-0. She improved slightly (is less passive). She speaks unconnectedly and laughs alone. At the current time, the patient almost does not speak and does not even respond to her name. She continues attending the Occupational Center and is able to eat by herself.

This would be a case of lost skills and regression (due to depression), coinciding with the rejection of the disability at the time of a change in her life: her brothers leaving home, which made her aware of her difficulties and her dependence.

Clinical case 3

A 25 year-old female who, after normal schooling in which she learned to read and write correctly and get by with mathematics satisfactorily within her disability, at 14 years-old met school friends with DS and reacted with rejection, although she later integrated well in a group. During adolescence she fell in love with an actor and spent hours watching videos that she searched for in the computer.

At 23 years she began to have "odd habits" (continually cleaning her teeth until her gums hurt) and complaining of a sore throat, until stopping eating. She finally had to be admitted, with a diagnosis of depression and medicated with fluoxetine.

She later stopped speaking and had an aggressive crisis that required another hospital admission. The family mentioned a deterioration in her cognitive skills. Months later, after several medical complications and serious problems, the anxiety and aggressive symptoms were accentuated, for which she was prescribed Risperdal® and Rivotril®. A reduction in anxiety was achieved, sleep was normalized, appetite increased, and she began to lose hair.

Despite experiencing a slight improvement, the patient never recovered her previous cognitive skills or achieved the level of independence she had before. She currently presents with selective mutism and refuses to go out of the house.

This would be a case of loss of functional and cognitive skills that also coincide with being aware of the disability. In this case an aggravation of the symptoms was caused to medical problems.

Discussion

The regression and loss of skills or abilities is observed in both depression and psychosis, and in the beginnings of neurological deterioration. The fact that it presents in young patients could lead us to rule out neurological aspects. In all the cases that we have seen there is a depressive base, motor slowing down and some manifestation of an obsessive disorder. Transitions and becoming aware of the disability, together with significant life events, and poorly prepared, are the common denominators.

Being aware of the DS, the assimilation and acceptance of the disability, together with the discovery of multiple skills favor the construction of identity[12]. Identity refers to the group of body, mental and psychological traits that are developed throughout life and that configure the personality. The construction of identity is a very complex phenomenon

that is formed using personal experiences and one's image perceived in others. Individuals with DS may experience sufficient emotional well-being to confront the adult phase of their lives and, as a result, join the working world and adult society where they have the possibility of working on their identity, knowledge of themselves, the discovery of their skills and their difficulty in being accepted as they are, and construct a self-esteem nourished on real facts[13]. To prevent this type of pathology —regressions and depressions— it is fundamental to work on the identity and the acceptance of the disability from early ages.

Adolescence involves an identity crisis that can be very intense in individuals with DS. As we have seen in the cases presented, regression has been observed at this age or in early youth. Regression, as a symptom, is very difficult to observe at first, thus it is very important to perform neuropsychological examinations from 18 years-old and to work on, from a very young age, the identity and acceptance of the ability.

To help a child to see their skills, as well as to understand what the disability does not involve, to boost their skills and to discover their abilities, without denying that there is much work to do, will help them to grow up harmoniously.

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