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Chronic Psychopathological Disorders and Down Syndrome

Giovanni Maria Guazzo* and Consiglia Nappo

IRFRI-Gruppo Forte, Salerno, Italy.

*Correspondence:
Giovanni Maria Guazzo, IRFRI-Gruppo Forte, Salerno, Italy.

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ABSTRACT

Down syndrome is the most common genetic disability. Their life expectancy has increased dramatically in recent years, probably because treatment and quality of life have improved. Still, despite this, the risks of developing chronic psychopathological disorders (including anxiety and phobias, depression, eating disorders, and autism) in adolescence and adulthood have increased.

In this paper, the differences in the presentation of the various psychopathological disorders in the syndrome are discussed, together with the strategies most frequently used to improve mental health in people with Down syndrome.

Keywords
Down Syndrome, Psychopathology, Anxiety, Fears and Phobias, Mental Health.

Introduction

Down syndrome is the most frequent autosomal trisomy (chromosome 21- C21) occurring naturally in about 1 in 700 live births. The prevalence rate of intellectual disability in the general population is estimated at between 1 and 3%. Chromosomal abnormalities (both numerical and structural) are responsible for up to 28% of intellectual disabilities, within which aneuploids are well represented. As the name suggests, what biologically defines Trisomy 21 is the presence of three copies of chromosome 21, instead of the usual two, in the body's cells. This excessive genetic richness results in abnormal development of the foetus and child, with consequences that persist throughout life. Human chromosome 21, technically called Hsa21, harbours around 250 protein-coding genes (the precise number varies depending on the genome annotation) and between 165 and 404 non-coding RNA (ribonucleic acid) genes that regulate gene expression.

Overexpression of proteins, linked to gene triplication, results in abnormalities involving the subject's heart, nervous system and gastrointestinal tract. However, almost every aspect of the Down syndrome phenotype is subject to an essential degree of inter-individual variability due to its polygenic nature and interactions with environmental factors. It occurs in several forms [1].

The most common (95% of cases) is the standard, complete and homogeneous trisomy (karyotype 47 + 21), which is characterised by a triplication of Hsa21 in every cell of the body. In the free mosaic trisomy (mosaicism; 1 to 2% of cases), on the other hand, only a proportion of the cells carry an extra Hsa21. The proportion depends on when the triplication of Hsa21 occurs (first, second or third cell division). Partial trisomy 21 (rare) defines cases in which only part of C21 is triplicated. Another possible cause of Down's syndrome, but infrequent (about 5%), is the Robertsonian translocation of part of chromosome 21, which can result in different chromosome formulae: 46 t (21; 21) + 21, 46 t (14; 21) + 21, and 46 t (15; 21) + 21, and 46 t (15; 21) + 21 (altogether 3% of cases). Partial trisomy 21 (less than 1% of cases) testifies that only one segment of Hsa21 is triplicated [2].

The steady increase in the life expectancy of people with Down Syndrome is impressive: it was only a few years at the beginning of the 20th century, 12 years in the 1940s and again around 30 years in 1970 [3]. The extraordinary improvement is attributable to greater care, particularly family care, for children and adolescents with this condition (in the first half of the previous century, they

were still more often abandoned in 'specialised' institutions), and better medical treatment. This vital progress already provides a good example of what can happen when living conditions are more appropriate.

The residual difference between life expectancy in Trisomy 21 and the general population can be explained, on the one hand, by the persistence of a still too high mortality rate in the early years of life, due to serious diseases (e.g. heart disease) that can sometimes be combined [4], and, on the other hand, by a higher premature mortality rate among older people with Alzheimer's [5].

In recent years, genetic research has certainly opened up fascinating avenues of knowledge. However, in the absence of effective therapeutic treatment, the only valid alternative is educational strategies and their ability to improve the functional level of the person with Trisomy 21. In this way, they will be able to improve their existence in many aspects and lead satisfying lives as adults. The ultimate educational-rehabilitation objective is to bring the person with Down syndrome to the maximum level of his or her potential to improve the quality of his or her existence. In this logic, much more can be done than what has been done so far (acquisition of cognitive, affective-emotional, social, and work skills) to improve the quality of life consistently [6].

As it is easy to guess, these aspects find their optimal place in the concept of 'Quality of Life'. The objective of quality of life, referred to each person, and therefore also to those with Down's syndrome, is realised in the search for the most significant possible development, both in terms of the acquisition of abilities and of affective-relational capacities and in the promotion of the broadest possible unfolding of these possibilities.

But what happens if the Down person presents some psychopathological problem which creates significant suffering and inability in social, work and other vital areas of life?

Psychopathological Disorders

Psychopathological phenomena, for reasons which are not easy to understand, have long been neglected in the study of intellectual disabilities, at least in part, as if they were of marginal importance for the quality of their lives. According to some [7,8], this disregard was determined by the prevailing interest of researchers and practitioners in the person's cognitive characteristics, learning skills, integration and inclusion processes, etc. In other words, it would have been a form of *overshadowing*, determined by the fact that intellectual disability is such a relevant characteristic that it 'obscures' the importance of emotional and affective, and more broadly psychopathological, disorders. The only exception to this situation is behavioural problems, for the simple fact that it is these that impose themselves, sometimes unpleasantly, on the attention of the researcher and the practitioner [9].

What do we know about the psychopathology associated with Down syndrome? Only recently has research been carried out in this respect; previously, not so much importance was attached to it. It was thought that people with Down syndrome only had intellectual problems and associated organic pathologies (hypotonia, heart disease, articulatory laxity, vision and hearing disorders, etc.). Unfortunately, it has been found that a person with Down's syndrome has about 30% more risk of psychopathologies, i.e. he or she is much more vulnerable than one would think. This may also be linked to the context in which he/she grows up: for example, if he/she lives in institutions and/or residential centres, the psychopathology, i.e. the most evident pathological problem, is linked to behavioural disorders such as self-stimulation. On the other hand, when the subject is integrated in different contexts, the most frequent pathologies are depression and anxiety [10].

Anxiety and depression syndrome, which tend to affect mainly people with Down's syndrome who are integrated into society, are, in all probability, the result of certain specific causes: 1) the comparison that the person with Down's Syndrome makes with others, pupils or colleagues, may lead him or her to ascertain his or her own greater difficulty in carrying out tasks, particularly the more demanding cognitive ones, and thus to confirm the perception of his or her disability (learned helplessness); 2) the consequent loss of self-esteem may dangerously lead this person to a gradual break in social ties and to a risky approach to depressive syndrome which, in this case, presents itself in an even heavier way than in other people [7].

Where, on the other hand, the person with Down's syndrome is not integrated into society, but lives most of his time in residential or quasi-residential structures, his psychopathological disorders can be traced back to 1) excessively rigid facility routines; 2) lack of professionalism on the part of educators and operators, who are rarely trained to deal with situations of acting out, outbursts of aggression or self-injury; 3) severity of cognitive impairment (people with severe and profound deficits are much more likely than those with mild and medium deficits to express themselves using 'self' and 'hetero-aggressive' behaviour).

These considerations make it clear that it is difficult for educators to implement a highly effective intervention programme. However, successes have been achieved through an integrated treatment of pharmacological and educational interventions [5].

At this point, it is inevitable to enter into the heart of the psychopathological dimension, presenting the problems which most frequently appear in the person with Down syndrome.

Anxieties, Fears, and Phobias

Anxieties, fears and phobias can be understood as the body's responses to any request for modification made on it. These physiological and behavioural responses are mediated by an emotional activation induced by the individual's cognitive evaluation of the stimulus. Anxiety, therefore, is an adaptive physiological reaction that, however, can take on pathological significance when it is produced too intensely over long periods or when it is hindered in its regular course [11,12]. It can be considered as a multidimensional construct with components: 1)

physiological: tachycardia, altered blood pressure values, profuse sweating, atypical body temperature, visceral contractions, irregular breathing, redness in the face, need to urinate, etc.; 2) *motor*: stereotyped and convulsive movements, tendency to move away from the anxiogenic situation (flight behaviour), trembling, immobility (freezing), tendency to avoid the anxiogenic situation (avoidance behaviour), etc.; 3) *verbal*: altered voice, stammering, interrupted speech, etc.; 4) *cognitive*: difficulty in paying attention, which is directed towards the individual's internal processes, which thus manages to escape from the anxiety situation (negative reinforcement); difficulty in retrieving information stored in the long-term memory, negative thoughts, etc. [9].

These behaviours not only involve the person as a whole, but also prompt him or her to flee from the anxiety situation and even to avoid it whenever possible.

The various studies conducted within this area show that around 35% of people with Down syndrome present problems related to anxiety, fear and phobias. In more detail, the feelings most frequently associated with Down's syndrome are those of personal inadequacy and inferiority, low self-esteem, hypersensitivity to evaluations expressed by others, etc. [8,13-15].

Depression

Of the various classification systems developed in recent times, we favour the binary 'unipolar-bipolar' system. In the case of the former symptomology, the person tends to break his relationships with the world, to lose all and any confidence in himself and the people around him. To these symptoms can be added others such as insomnia, loss of appetite, a protracted feeling of asthenia, etc. In the latter's case, others of the opposite sign are added to these symptoms. In other words, the person may cyclically present manic episodes, during which he shows a grandiose and completely unjustified self-esteem, exhibits a total inattention that prevents him from any form of concentration, etc. As far as the mentally retarded population is concerned, depressive syndrome (uni and bipolar) was only recognised in 1983 [16] and it is perhaps for this reason that we have epidemiological data (about 12% of subjects) that are still somewhat lacking for the population with Down's syndrome [17].

Finally, as far as the theories elaborated to identify the causes of depression in people with Down's syndrome are concerned, the one that has gained the most consensus was elaborated by Seligman [18] and goes by the name of 'learned helplessness'. It is characterised by the belief that the person gradually learns not to expect anything positive first from the world around him and then from himself, with the result that he is led to increasing psychological distress. In this case, the person with Down Syndrome is led to believe that failure is inevitable and that, consequently, nothing can be done either to cope with a positive situation or to avoid a negative one [19,20]. Learned helplessness leads to problems on a cognitive, emotional and motivational level. At the cognitive level, there is the perception of lack of control over the situation; at the emotional level, there is fear, dread and anxiety in the face of situations one does not want to face, depression at one's own inability to succeed and apathy and resignation in the face of failures, which are considered inevitable. At the motivational level, one tends to attribute the successes of one's actions to external causes (external *locus of control*), a lowering of self-confidence and self-esteem [9].

Learned helplessness would be acquired by individuals with Down syndrome as early as primary school and further consolidated in later years of schooling.

Eating Disorders

The literature on eating disorders in Down syndrome has mainly considered overweight and obesity, completely neglecting bulimia and mental anorexia, given their low incidence in the population. Instead, space was given to omnivagia and food refusal, which are predominantly present in people with severe and profound cognitive deficits.

A person is considered overweight when he or she exceeds his or her target body weight by between 10% and 20%. Beyond this percentage, the person is considered obese. This leads both to an increase in risk factors for cardiovascular, respiratory and orthopaedic diseases and the general state of well-being, and to a subjective deterioration of body image, which often has severe negative repercussions on the psychological side [7].

The causes of obesity are multiple, although the role played by the genetic component is evident. Bad eating habits often compound this, the use of food as a means of reducing anxiety, or something else.

About the incidence of obesity in the population with Down's syndrome, several studies [21] have shown that 37.14% of males are obese or overweight, compared with 34% of the normal population. The percentage rises to 38.46% when it comes to females with Down's syndrome, whose distance from the normal population (24%) is far greater than that of males. However, we can see how parents all too often use food, and sometimes by educators, to please the person with the syndrome and let him or her spend their free time, reducing the likelihood of behavioural problems appearing.

Autism

The diagnosis of autism is rather tricky and becomes even more uncertain in persons with Down syndrome or other known forms of intellectual disability. Several studies [22-28] have found that, although there is no epidemiological study of the prevalence of autistic-type disorders in a large population of children with Down Syndrome, what is usually evident is that the incidence is in the order of 10%. It isn't easy to have precise data, as many cases are not diagnosed, or are diagnosed at an advanced age. In the case of Down's syndrome, the parents of a child, and the carers themselves, are busy with the various clinical problems (heart disease, hypotonia, altered thyroid function, joint laxity, visual disturbances, etc.) typical of the syndrome and with the

multiple issues that the child suffers from Down's syndrome. Typical of the syndrome and with the various aspects of the expected developmental delay (deficits in gross and fine-motor, communicative, sensory, performance skills, etc.) and, precisely because of this expected delay, the possibility that autism may also be present is not considered. Moreover, many of the typical symptoms of autism can be regarded as characteristic of the syndrome and not sufficient to diagnose the disorder [29-31].

The area that is most impaired during the development of the child with Down syndrome is the cognitive area, i.e., the development of thinking, reasoning, and comprehension, and a delay is expected in this area of development. Another area of development is the socio-emotional one: it has a more normal development in a child with Down's syndrome than in an autistic child [32,33]. Thus, children with Down's syndrome in whom one suspects (or to whom one can add the diagnosis of autism), there are behavioural disorders, typical of autistic disorder, generally show deficits in the following areas [33]:

a) High degree of isolation: The child does not usually relate to others and prefers being left alone. He seems to regard people as objects, not human beings. He does not join playgroups with other children, and, in contrast to those who only have Down Syndrome, who are generally very affectionate and expansive, the child who also has autism often does not want to be hugged;
b) An anxious and obsessive desire to preserve one's status: any change in the daily routine can cause great discontent;
c) Lack of eye contact: it is typical of autistic people not to have eye contact with their interlocutor;
d) The presence of repetitive and 'stereotyped' motor behaviour: e.g., sitting for long periods with an object in one's hand, moving it back and forth, and staring at it.

In addition to these problems, which are present in almost all individuals with Down syndrome, there are others which are of greater concern to parents and other caregivers, and these are the 'dysfunctional behaviours' which, because of their intrusiveness and dramatic nature, are dealt with separately.

Dysfunctional Behaviours

Dysfunctional behaviours belong to a class of behaviours that are not considered relevant to the context in which the individual is found. Generally, this behaviour class interferes with the person's learning and socialisation process, creating uncomfortable situations. However, this hindering and interfering role towards development is not always well understood, especially when placed within a proper psychopathological view.

This model predicts that the stabilisation of maladaptive responses may occur more rapidly than development, through a series of stages. By this model, certain types of apparently problematic behaviour may initially develop in a particular context and with certain contingencies and later be maintained due to associated side effects. According to this approach, severe problem behaviour develops through gradual shaping of responses, mainly through social contingencies. The assessment, therefore, should focus on identification and maintenance factors: 1) identifying early types of behaviour that share similar aspects with severe problem behaviour, but which may be of lesser intensity and without causing, for example, tissue injury (lightly banging one's head on objects, biting without leaving teeth marks, scratching without necessarily tearing the skin, pulling one's hair, etc.), called 'pre-diadaptive behaviour', and 2) demonstrating that these acts are linked to environmental contingencies [34].

Among the various inappropriate behaviours that a person with Down syndrome may adopt, the aggressive response is undoubtedly one of those manifestations that arouse the most discomfort and concern in caregivers. Such a response consists of a series of behaviours intentionally aimed at producing harm to persons, things and structures. This modality implies that a behaviour can only be considered aggressive if regarded as an intentional expression. The problem, at this point, is: 'Does a severely Down person want (i.e. is there intentionality) to produce harm?'. It is necessary to determine if such intentionality exists; the subject is aggressive. If it does not exist, then it is simply harmful behaviour. He who intends to' use his head' to produce harm, and who does not have it responds automatically. How does one assess the aggressiveness of a person with an intellectual disability? One has to use a card for the systematic observation of behaviour or 'functional analysis', i.e. to see how often the subject is aggressive, with whom. In these contexts, when performing which tasks [35].

Only in this way can it be discovered that the person is often aggressive or harmful (damaging), depending on our behaviour. The analysis of what people do with those who are aggressive is critical. In fact, except for a few cases where harmful behaviour is maximised, the person in a situation of disability only affects certain people and not others. The sign of fear in those around him can be a discriminating stimulus to evoke an aggressive response. So, what do you do when a person is aggressive? It depends, first of all, on the age. Again, techniques are based on reinforcing the person, taking away attention when he is aggressive, and using 'occupational activities' to manage the tension that triggers aggression.

For example, occupational activities teach valuable things regarding *existential functioning*, and situations that encourage 'choleric' behaviour are avoided. One cannot teach an aggressive person not to be assertive by attacking him; it is a contradiction. Instead, he must be taught that other methods can be used to satisfy needs. It is the golden rule of interpersonal relations: if I want something, I must be willing to give something in return. In educational terms, we can teach him this and modify his strategies.

Intervention Strategies

Intervention on the Down person with psychopathological problems does not aim at the simple extinction or decrease of maladaptive behaviour. Still, learning situations are organised in such a way as to provide him with positive emotional experiences. In this way, in addition to fostering new learning, the child is allowed to improve

his or her self-esteem and decrease the performance anxiety that often prevents him or her from tackling new situations. Moreover, the new skills learnt are always aimed at learning increasingly complex and refined skills that converge to make the Down person achieve the most significant possible autonomy and foster the adaptive process.

The available intervention strategies are relatively numerous and each of them has characteristics, advantages and limits which make it preferable in some cases rather than in others, as one can get an idea from the following descriptions even if they are not exhaustive (the reader is referred to the reading of [35,36], for valuable in-depth studies) (Figure 1):

Intervention package A: *Anxieties, fears and phobias*. The terms anxiety, fear and phobia are not clearly distinguished, but are used to denote the same theoretical construct. Fear, however, seems to refer to particularly violent tensions, which generally emerge about circumscribed situations (fear of examinations, fear of public speaking, etc.). A fear reaction comprises three components: 1) a state of high tension, 2) a state of activation of the autonomic nervous system and 3) flight and avoidance behaviour. These components are autonomous, i.e. they can vary independently of each other. In humans, as in animals, fear responses that cannot be traced back to specific stimulus-situations can also be observed; for example, novel and very intense stimuli, in addition to eliciting specific orientation responses, can induce

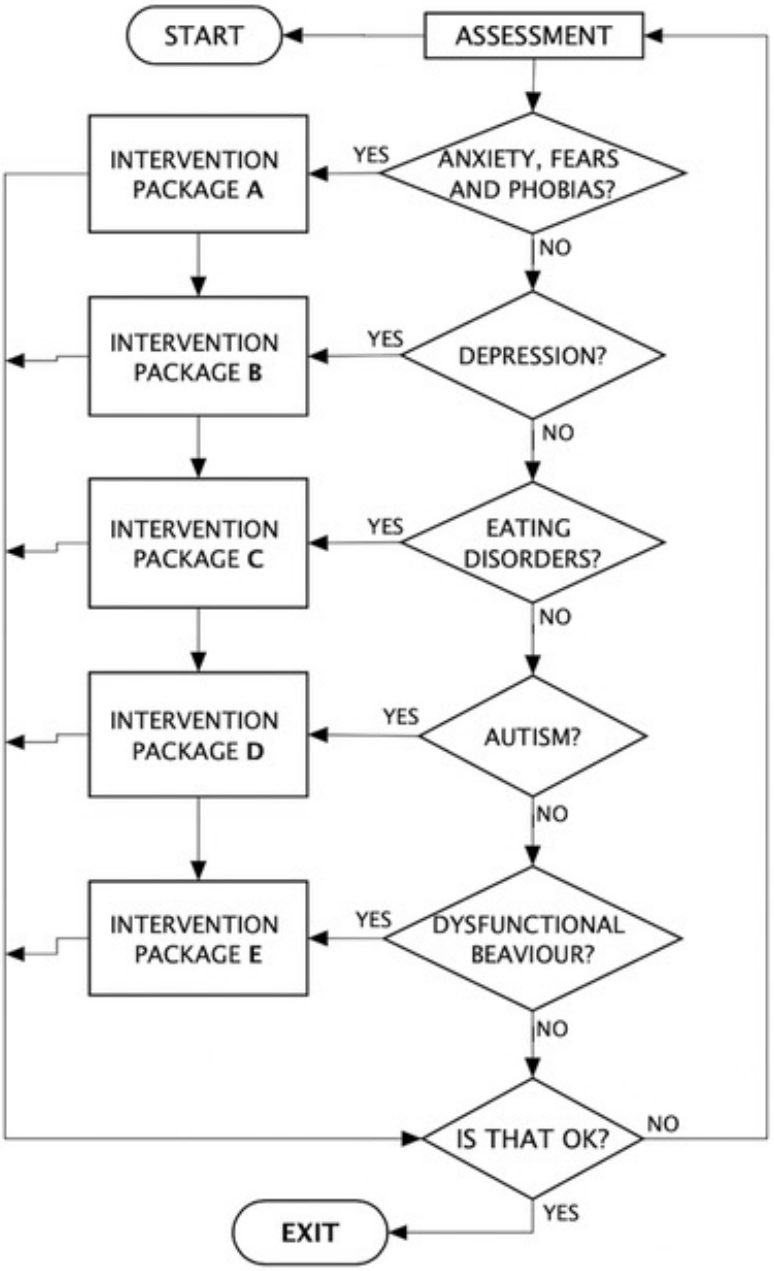

Figure 1: Flow-chart relating to the choice of strategies best suited to individual psychopathological disorders.

fear responses. However, most fears, at least in humans, originate in precise learning processes [37]: classical conditioning, operant conditioning and shaping.

Very intense and persistent fears can give rise to phobias, which are characterised by the presence of the following factors: 1) disproportionate reaction to the situation; 2) control impossible through rational analysis; 3) avoidance of the feared situation; 4) voluntary control impossible; 5) permanence over a relatively long period; 6) maladaptation; 7) lack of any connection with ontogenetic developmental stages.

These characteristics differ only quantitatively from the physiological and avoidance responses in subjects afraid in certain situations. Moreover, phobias, like fears, are also learnt according to classical conditioning, operant conditioning and modelling principles. However, the most interesting aspect that characterises phobic behaviour is its resistance to extinction, i.e., it persists, even when the individual no longer comes into contact with the event that induced the phobia. This occurs, most probably, because the phobic is driven to emit avoidance behaviour, the emission of which reduces the anxiety present in the organism (negative reinforcer). The individual will tend to avoid the feared situation [38] constantly.

Thus, classical conditioning is responsible for learning phobias, while operant conditioning is responsible for their maintenance: avoidance inhibits extinction [39]. Fear learnt according to the principles of classical conditioning tends to be maintained through the intervention of operant processes: the child, in the presence of anxiogenic stimuli, emits those behaviours that distance it from the feared situation. This implies that the choice of intervention strategies is subordinate to how the fear and/or phobia is learned: e.g. *counter conditioning* for fear and *systematic desensitisation* for phobias, etc. However, the child's development and adaptation should never be subordinated to a procedure: if it does not promote general improvement or is 'harmful', it should not be used.

All strategies used for the control of anxiety, fear and phobias are based on a direct application of data acquired from experimental psychology and refer to three different procedural phases. The first phase, in the intervention to reduce and possibly eliminate excess anxiety, as with any psychological intervention, consists of a precise description of the anxious and phobic behaviours: they must be identified and described in such a way that all those working with the subject know how to recognise them when they are to be observed. The second phase consists of observation of the anxious and/or phobic behaviour, preferably in functional analysis. The third phase involves relaxation training aimed at achieving a state of mental tranquillity and muscular relaxation. The training consists of putting various muscle groups under maximum tension and then relaxing them completely [40]. Muscle relaxation has no particular contraindications. In the worst-case scenario, the person may experience initial difficulties producing relaxation. These, however, tend to disappear in a short time. Obstacles may arise when the person cannot exert pressure on specific muscle groups or discriminate between tension and relaxation. This difficulty is widespread in children and pre-adolescents, for whom relaxation may present problems of a cognitive nature. In this case, it is advisable to resort to playful activities in which the child can selectively exercise different muscle groups: for example, having the child mime the kangaroo jump over small obstacles or the cat position. In addition to muscle relaxation, breathing techniques, mindfulness, listening to music, dancing, etc., can also be used.

Intervention package B: *Depression*. As we have seen, learned helplessness is one of the main reasons for depression in people with Down syndrome who, after being exposed to frequent aversive stimuli, become unable to avoid the subsequent encounter with these same stimuli, even if they are avoidable. This presumably happens because they have learned that, despite their best efforts, they cannot control the situation.

What then can be done to overcome learned helplessness? First, it is essential to recognise and change the negative thoughts, observing them and identifying when they start to occur, that feed this state and replace them with more positive and realistic ones. Furthermore, it is helpful to focus on what is within one's control, set small, easily attainable goals and reward oneself for small successes, and then move on to more ambitious goals; instead of focusing on what cannot be controlled, focus on what can be done to improve the situation. Other effective strategies are: creating a positive social environment (surround yourself with encouraging and supportive people, and participate in activities that help you socialise) and cultivating your self-esteem (identify your positive aspects and value them, look at the past not as a series of failures, but as a learning opportunity, do not be too harsh on yourself).

Intervention package C: *Eating disorders*. In both sexes with Down's syndrome, excess weight is often detected and this is extremely worrying as it can aggravate the consequences of specific pathologies associated with the syndrome (respiratory diseases, congenital heart disease, joint problems, etc.), as well as adversely affecting the development and performance of the most common motor activities. Based on these considerations and beyond the medical explanations on slowed metabolism, altered leptin concentrations and reduced physical activity (even if, despite practising sport, the body mass index remains, in most cases, high), it is opportune to direct intervention towards simple formulas of self-regulation. One of the most frequently used strategies that has shown a tendency towards weight loss, albeit not a resounding one, has been the 'educational contract' [41], a procedure in which both the required behaviours or tasks and the reinforcers that follow the appearance of those behaviours are specified [42].

The essential points needed to draw up a good educational contract are: 1) establishing exactly which activities are to be rewarded with tokens (symbolic reinforcers: dots, colours, ribbons, etc.), 2) specifying the reinforcers to be used for the tasks to be rewarded, 3) specifying the reinforcers to be used for the tasks to be rewarded, 4) specifying the tasks to be rewarded and the reinforcers to be used

for the tasks to be rewarded.); 2) specify the reinforcers the subject will receive if he/she behaves in the desired manner; 3) specify the amount of points he/she will lose if he/she misbehaves; 4) list the rewards the subject will be able to obtain thanks to the symbolic reinforcers; 5) determine when the exchange of the tokens with the rewards can take place; 6) record the learner's behaviour. In other words, it is essential to: a) make a list of all the rewards that can be exchanged with the symbolic reinforcers; b) determine how much each reward will cost (obviously the more coveted the reward, the more expensive it will be); c) determine when the exchange between the symbolic and the support reinforcers will take place; d) use the response cost if the subject displays behaviour that is difficult to manage. In such a case, the educator: a) decides on and describes some problem behaviour that causes the subject to lose points; b) specifies how many points the subject loses each time he/she misbehaves. In any case, the number of points the child loses must be low; excessive loss risks demotivating the subject; c) when the problem behaviour manifests itself, he/she removes the symbolic reinforcers without comment and aversive stimuli [35].

Intervention package D: Autism. The same strategies apply to this package as with autism spectrum disorder and refer to the principles, procedures and interventions from the field of behavioural psychology (Applied Behaviour Analysis). The effectiveness and speed of learning depend on the teaching procedures used to achieve the formulated educational goals. These procedures must be coherent (with the educational objectives being pursued) and stimulating (persistence in the task). Still, they must also be selective (priority tasks for the learner), effective (way of organising and presenting tasks) and contingent (reinforcements that follow some responses and not others). They must also guarantee the learner certain general principles:

- a criterial evaluation of the skills mastered;
- a continuous monitoring of progress, objectively measured;
- the visualisation of the learning 'curve';
- an adequate mastery criterion;
- the structuring in hierarchical sequences of the objectives to be pursued (task-analysis);
- a functional time scansion to achieve the mastery criteria for each performance;
- adequate learning opportunities;
- educational methods to increase the frequency of correct answers (reinforcements and reinforcement programmes); - individualised and group learning moments when the pace of learning requires it.

These principles are fulfilled in all the teaching programmes inspired by ABA: *Discrete Trial Training*, *Natural Environmental teaching*, *Verbal Behaviour Teaching* and Functional *Communication, etc*. [36], and which are effective in the field of learning. [36], and which are effective for people with Down syndrome and autism.

Intervention Package E: Dysfunctional Behaviours. How can these problems be solved? One answer, which is general but decisive for improving the quality of life of the person with disabilities, is to involve them in activities, preventing them from being deprived of stimuli. A fundamental concept is that every person with atypical development can learn something, however small. Research done on very primitive animals, such as pigeons, has shown that these, if properly trained, can do incredible things [43]. So if you can teach pigeons, you can certainly teach more sophisticated stuff to people with a much more evolved cerebral cortex.

Once you have established a valid hypothesis for a given individual, the intervention is relatively standardised. First, a baseline is created, i.e. one observes how often the subject behaves dysfunctionally [44]. Then, one has to record how the problem usually occurs, where it occurs, in which context and next to whom. It has been noted that inappropriate behaviour is not constant, i.e., the subject will be more aggressive with certain operators, in particular situations, or due to other environmental circumstances. Suppose one is a careful observer of this development. In that case, one can already make hypotheses (perhaps the relationship with the operator creates anxiety to which the subject responds through aggression). Having ascertained this, one must choose the various intervention techniques (of which there are few). A practical method is based on 'differential reinforcement' (do not give attention when the subject self-harms, or attacks others, but give it when he/she misbehaves (i.e. attention must be directed differently).

It has been noted that this phenomenon decreases in many cases when appropriate behaviour is reinforced. To achieve this, however, there is a big problem: one should ensure that everyone around the person does the same. If the person is placed in a community (school, socio-educational community) there must be behavioural consistency. Planning must be done with several people who share that objective and methodology. Failure to do so produces a worse response than the one from which one started. Differential reinforcement can be done in many different ways [35].

Another technique is *communicational training*, the purpose of which is to lead the subject to avoid situations that evoke maladaptive behaviour and to use other communicative modalities that are functionally equivalent to those that are dysfunctional in saying 'NO': one must, for example, teach him/her to shake his/her head in a meaningful way to communicate 'NO'. In this way, the subject increases his or her communicative capacity and, thanks to this, can manage the relationship with the other, thus becoming a little freer than in different situations without engaging in aggressive behaviour [45].

Conclusions

The person *model*, taken into consideration, is a very integrated system between the cognitive repertoire (totality of the knowledge a person uses to interpret and interact with the world), the affective-sentimental repertoire (how one lives specific experiences or what are the affective disturbances linked to feeling) and finally the socio-relational repertoire (how to interact with the other). These three elements are closely related, i.e. it only takes gaps in one or the other of these three elements (pieces of the person) to create

difficulties in the learning process.

What happens when the person, for reasons that may be linked to a disability or performance deficit or a socio-cultural disadvantage, has a chronic psychopathological disorder and is not ready to express themselves linguistically as society requires? What could be the consequences on an affective, cognitive and relational level?

The first fact is that the individual (child or adult) lacks units of information; when others speak, he cannot understand what is being said and reacts by 'closing in', isolating himself, debasing himself, and even adopting feelings of inferiority. This undermines the sense of self-esteem. A cognitive problem has immediate negative consequences on the affective, emotional, and social-relational levels. A person who feels inadequate in some way to the demands of the environment is very likely to adopt two relational strategies: 1) aggression, which is the result of frustration ('... I feel inadequate so I react to eliminate, in a purely imaginative way, the source of my disorder') and 2) passivity, which is the result of social inadequacy ('others do not seek me out, I keep to myself'). This happens with many people with intellectual disabilities who feel '*cut off*' if their interlocutor does not use the same linguistic code as them [46].

The intervention, then, must be personalised, prolonged in time, and diversified and integrated. A fundamental aspect is the involvement of both parents and the collaboration of all the educational agencies that revolve around the person with Down syndrome. As we have seen, one treatment that fully meets these requirements is behavioural.

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