# Computer-Based Cognitive Training in Adults with Down's Syndrome

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**Abstract.** Adults with Down Syndrome show a clear genetic susceptibility to developing Alzheimer's Disease, the most common cause of dementia worldwide. In this paper we describe a set of computer-based exercises designed for cognitive training of adults with Down Syndrome. The aim is to provide telerehabilitation via a Web application that can be used at home to create an enriched environment. Each exercise is presented as a game with images, text and vocal communication. The user moves forward at increasing levels of difficulty according to previous positive percentage thresholds. Performance data is centrally collected and available to the tutor to check progress and better define the training. Several categories of exercises are needed to train different abilities: attention, memory, visual-spatial orientation, temporal orientation, prelogical and logical operations, perception, visual analysis, language, and data relevance. At this time, two modules have been implemented for exercising attention and memory.

**Keywords:** Training software, tele-rehabilitation, Down Syndrome, dementia, accessibility, learning games.

## 1 Introduction

Down Syndrome (DS) is the most common form of mental retardation of genetic origin. In the last few decades, life expectancy in people affected by DS has increased greatly, from an average of 25 years of age in the 1980s to an average age of 60 years [1, 2]. In Italy, about 60% of all subjects with DS are adults [3].

Adults with DS show a clear genetic susceptibility to developing Alzheimer's Disease (AD), the most common cause of dementia worldwide. Neuropathology of AD can be observed virtually in all subjects with DS older than 40 years of age, while the onset of the clinical signs of dementia is on average observed around 50 years of age [4].

Strong evidence supports the beneficial effects of enriched environment (EE) on several aspects of brain development and brain plasticity in the mouse model of DS [5]. Preliminary results of a randomized controlled ongoing trial at IRCCS Stella Maris have assessed the effect of EE intervention based on psycho-physical training for the prevention of functional and cognitive decline in adults with DS.

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In this paper we describe a set of computer-based exercises designed for cognitive training of adults with DS, called “STELLA DS” - Software To Exercise Learning and Language in Adults with Down Syndrome. The aim is to provide tele-rehabilitation through a Web application that users with DS can utilize at home to create an enriched environment to train the brain and prevent dementia.

Each exercise is presented as a game with images, text and vocal communication, e.g., statements, cues, and reinforcers. The user moves forward in increasing levels of difficulty according to positive results, assessed by percentage thresholds; otherwise he/she remains at the same level or goes back. Tutors can personalize the training depending on the skills and preferences of each subject. Performance data for each subject and exercise is centrally collected and available to the tutor for checking progress and issues, and to better define the training.

Several categories of exercises are needed to train different abilities: attention, memory, visual-spatial orientation, temporal orientation, pre-logical and logical operations, perception, visual analysis, language and data relevance. At this time, the first two modules have been implemented for exercising attention and memory. The effects of the use of the cognitive training through STELLA DS will be tested following a neuropsychological protocol that will check the degree of the enrolled subject’s abilities before and after the training.

## 2 Related Work

Tele-rehabilitation offers cost-effective services that can improve access to care for patients with neurological disorders. The techniques of tele-rehabilitation are now widely used internationally, and are based on online rehabilitative exercise monitoring by specialized medical personnel. The Internet offers new methods of intervention and a greater and more effective clinical supply. Tele-rehabilitation maximizes the potential of rehabilitation, allowing the clinician to assign specific tasks to be carried out daily at a distance, while maintaining full control and constant monitoring of the rehabilitation.

Nouchi et al. (2012) investigated the impact of playing brain training games for 4 weeks, in elderly subjects. Results showed improvement in cognitive function (executive function and processing speed). Results indicated that the elderly can improve both executive functions and processing speed in short-term training [6].

Using computerized versions of training programs has allowed the implementation of adaptive algorithms that ensure that the level of task difficulty is always challenging for the individual, something shown to be crucial for the training to be effective [7].

Computerized training has been shown to improve working memory performance in healthy children and adults [8-11], in adults recovering from strokes and other acquired brain injuries [12, 13], and in children born preterm [14], in children with attention-deficit hyperactivity disorder (ADHD) [15], and in children with intellectual disabilities [16].

Previous research in children with DS has indicated that rehearsal training can improve working memory [17, 18].

A recent study by Bennet et al. 2013 [17] evaluated the impact of a computerized visual-spatial memory training intervention on memory and behavioral skills in 21 children with DS, aged 7-12. Following training, performance on trained and non-trained visual-spatial short-term memory tasks was significantly enhanced for children in the intervention group. This study was carried out in school. A previous study [18], not based on computerized training, suggested that parents can be good trainers and auditory working memory can improve with parents as trainers.

There are no studies with computerized training in adults with DS to prevent the development of Alzheimer's Disease (AD). Strong evidence supports the beneficial effects of enriched environment (EE) on several aspects of brain development and brain plasticity. We aimed to study the improvement of cognitive abilities also in older DS subjects, with computerized training that can also be used at home with clinical monitoring.

Our application, unlike other software in use (such as Cogmed JM), is open source and free, relieving families and rehabilitation centers of the burden of license costs. Furthermore, STELLA DS do not require the user to install any software: it is a Web application, thus usable through a browser on any device connected to the Internet.

## 3 Designing the Software

As previously mentioned, there is evidence that supports the beneficial effects of EE on several aspects of brain development and brain plasticity in a mouse model of DS [5]. Preliminary results of a randomized controlled trial in IRCCS Stella Maris has assessed the effects of EE intervention based on psycho-physical training for the prevention of functional and cognitive decline in adults with DS. Based on this observation, it is crucial to maximize the time that the subject devotes to the rehabilitation. Moving from the classic face-to-face approach to a technology-enhanced therapy (i.e. executed interacting with a computer or a tablet) is a natural evolution today. Removing the constraint of physically obliging the DS subject to go to medical labs for therapy alleviates the burden on families, allowing subjects to exercise their brain more comfortably at home, independently and at their own pace. In fact, independence may encourage the subject to increase training time, while the electronic tool may provide more agreeable stimuli, both leading to better results. Tele-rehabilitation is conceived as a tool that enables the individual's independence in performing the training and thus empowers the DS person, taking full advantage of her/his residual abilities.

The STELLA DS software has been designed following participatory design principles, involving psychologists and doctors from the earliest steps of the projects, to guarantee the creation of a flexible and usable product that is pleasant for the users. Specifically, face-to-face meetings, Skype sessions, and interaction via email have been adopted as collaboration mechanisms, to collect user requirements and collaboratively design the software and its function. In order to achieve targeted features we defined main software design constraints:

- To propose the exercises as a serious game, with increasing/decreasing levels of difficulty, in order to dynamically adapt the trial flow to the subject's responses and interactions. This adapts the proposed trial to the pace of the student, decreasing stress and favoring the independence of the subject in carrying out the rehabilitation.
- To provide psychologists and doctors with a tool for easily assessing the learning trend of their patients and refining the training program.

The W3C Web Accessibility Guidelines (WCAG 2.0) and usability criteria drove the design of the software STELLA DS. The software exploits all the user's sensorial channels in order to provide an enriched environment that favors the subject's comprehension and stimulates attention, providing visual, audio and vocal information. For instance, to keep the subjects' attention, textual commands are shown on the screen and vocally announced. To sustain learning, a short demo video is shown before the game starts for teaching by visual examples. Furthermore, the touchscreen device allows one to use pointing, which may be more natural, intuitive and easy to use than the mouse and touch pad.

The system is accessible after a log-in phase. For DS people who may have difficulty remembering passwords, the log-in phase is simplified, requiring only a nickname. No time constraints are applied when the tasks are assigned to the student and (s)he can stop and resume the game anytime. Feedback and reinforcers appear on the screen during the exercise, and are also announced, to increase the probability that the subject manifests positive behaviors in the future. Pre-recorded audio files, instead of a speech synthesis engine, have been used to make interaction more comfortable for the user. After the development of the prototype, a pilot test with two users was carried out, to better understand the needs of subjects with DS.

## 3.1 The Architecture

The architecture consists of a web server integrated with a PHP interpreter and a MySql database. The classic LAMP Server -- Linux, Apache, MySql and PHP has been adopted. The database is used for data gathering, for adaptation, keeping the functional parameters, and for user management. It also enables multilinguism: depending on the selected language, a corresponding set of labels is visualized in the user interfaces (UIs) of the application.

To make the software (SW) fully flexible and configurable, each training exercise is implemented as a finite-state machine, defined through two database tables. One describes the states associated with the implemented page, the other the transactions from one state to another, based on the results of the previous state execution. In this way it is simple to modify the behavior of each module by only changing its description tables; no changes are required to the code.

The UIs are implemented with HTML 5, style sheets (CSS) and Javascript (specifically jQuery). Each user interface is controlled by a routine Javascript with Ajax calls, both for the content processing and their presentation. Through Javascript, the user input is controlled both for users interacting via keyboard, mouse/touchpad and touchscreen devices. The global architecture of the STELLA DS application is synthesized in Fig. 1.

Three kinds of users can interact with the system in different places and times, as shown in Fig. 1: a) Tutors (psychologists, doctors, parents and caregivers); b) Students/patients (adults with DS), and c) Administrator, the person in charge of changing system configurations and updating the software. The application is able to execute three main functions:

- Enable tutors/teachers/caregivers to assign one or more educational training modules
- Enable patients to carry out their assigned exercises/games. The exercise can be stopped and resumed at the next login, without losing data and maintaining the last state (ensuring the consistency of the intervention)
- Gather and process data related to the exercises executed (learning data analytics). Data is collected during the execution of Modules to assess the level of performance achieved. For each learning module a success rate has been defined in order to make the student progress through the levels. The tutor can monitor the performance data and decide to suspend the execution of a module when the level seems to be too difficult.

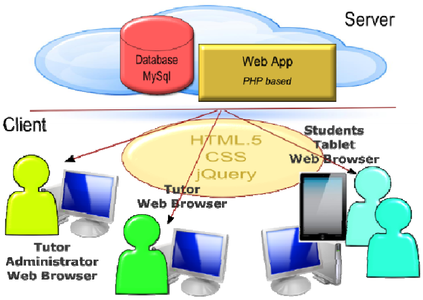


**Fig. 1.** Software Architecture

# 4 Software Components

## 4.1 Access Component

The access page represents the entry point of the application and requires a log-in. Depending on the user -- Administrator, Tutor or Student, the access component visualizes the appropriate user interface (UI) as defined in the database. In the following the different UIs are described.

## 4.2 Administration Tools

With the administration tools it is possible to create, modify and delete users, and set up values of the application parameters. Tutors and administrators can create "student" users by personalizing the profile according to several attributes, such as the speaker voice -- male or female -- and the language. This allows tutors to have more flexibility in profiling the user, adapting the exercises to their preference. After the creation of a student, the tutor may assign him/her a sequence of training modules to do at home, monitoring their execution and the performance data at any time. It is possible to cancel a student – a logical and not a physical cancellation -- to assure the integrity of the historical data stored in the database.

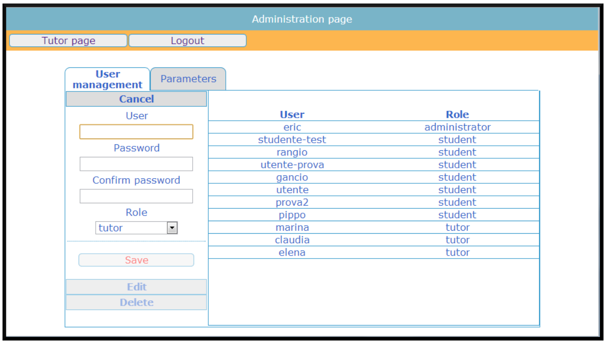


**Fig. 2.** Administration Tools

### 4.3 Tutor Interfaces

The tutor interface allows to assign one or more modules to a student. After login, the student will begin the execution of assigned modules in sequence (Fig. 3). Information on the exercises performed is kept in the database. The tutor interface collects and shows this information. A tutor can monitor the exercise the student is engaged in and possibly suspend the execution of a problematic one.

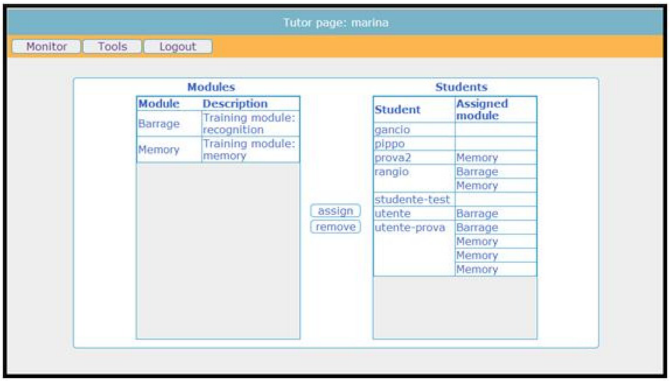


**Fig. 3.** Tutor Interface

## 4.4 Data & Statistics

During the execution of training modules, the data collected is used in assessing the level of performance achieved. This data is processed and shown with the details of each session and the possibility of obtaining a broader or in-depth view. For each learning module a simple statistical index has been defined that is referred to as the success rate characteristic of that specific module. An example of statistics is shown in Fig. 4:

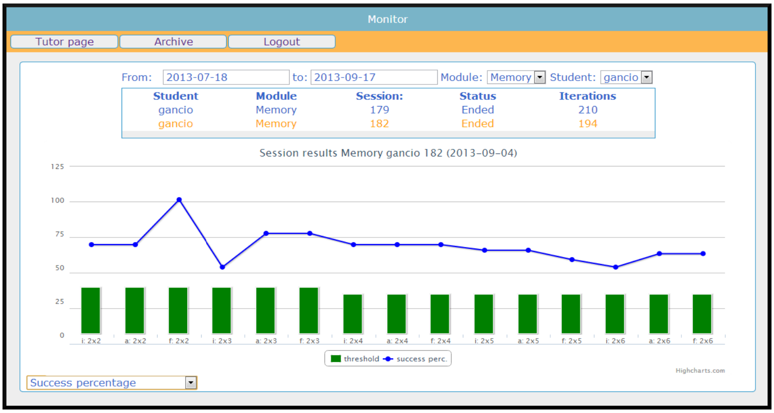


**Fig. 4.** Data & Statistics

## 4.5 The Student User Interfaces: The Learning Modules

This component implements the training exercises assigned to the students. The UI that implements the exercises is presented as a gaming session where student interaction occurs via mouse or touchscreen. JavaScript features have been used for the interactivity such as: animations based on the visibility and size of html page portions, use of timers to put the correct chronological sequence the actions, the control of user input, etc. The pages are scalable as much as possible to the size of the browser window, recalculating at each event 'window resizing' the optimal page view, in accordance with its specific geometry.

The task to be executed by the student is written in the upper part of the interface and also announced. In order to make interaction more comfortable for the user with a voice that is “warmer" and less "robotic" than the speech synthesis one, we used recorded voices. There are two possible voices: a male and a female have recorded all the words and phrases used in the UI and all data is memorized in the database. Javascript procedures reconstruct the phrase in the correct sequence, by combining, through the use of HTML 5 Audio objects, the pre-recorded vocal files. There can be multiple speakers for each supported language. The training exercises (learning modules) are the core of the application; thus, in the following we will describe them in detail.

## 5 Training Exercises

At this time, the first two learning modules have been implemented: 1) barrage and 2) memory. Each exercise starts with a demo that visually presents the assigned task, in order to better allow the student to understand the task and imitate the actions. The execution of each module can be suspended by the student, i.e., interrupted and resumed later at the same point where the user was working at the time of the stop.

### 5.1 Barrage

The barrage is specifically used for training visual attention and memory. The student is required to select "target objects" in a grid containing various elements, including distractors (Fig. 5). Target objects to be identified are listed by name in the upper part of the page; an essential and very strict (for clarity) command: Touch 'element lists' are visualized and also vocally announced. The objects are grouped in categories (objects, animals, numbers, letters, shapes) and the student has to touch it/them via the mouse or touchscreen (depending on the device utilized).


**Fig. 5.** Barrage UI

The number of target objects varies from 1 to 3, while the grids are composed of 3, 6 or 12 elements. The level of difficulty increases from the simplest (touch 1 target in a grid of 3 items), the less simple (touch 1 target in a grid of 6 items, and then touch 1 target in a grid of 12 items), to the hardest (touch 3 targets in a grid of 12 items).

This sequence of exercises is applied to all four identified categories, which are also of increasing difficulty themselves: 1. objects/animals, 2. numbers, 3. letters and 4. shapes. The whole schema of levels above described is first proposed on the first category, and then on the second and so on.

Every click of the mouse or finger pressure (click) on the touchscreen has an immediate feedback: a green tick, if the touch is correct, i.e., a target has been identified, or red "X" in case of error. The success rate for a level is considered the percentage of the number of correct answers provided by the subject compared to the total number of possible correct answers (< = 1). There is a tolerance threshold of the number of possible errors (we set 20%, but this parameter is configurable). If the number of errors goes above this threshold, there is a repetition of the trial in the same configuration. Two consecutive errors make the game return to the previous level of difficulty.

The user can explicitly ask for help (by pressing an icon), or otherwise help is automatically provided if, after a fixed time (configurable as system parameter by the tutors) the user has failed to solve the trial. A request for help leads to repeating the exercise, if the student does not provide the correct answers with a percentage of at least 60%. The help functionality automatically runs after a fixed (configured in the application's parameters) amount of time if no actions in the Barrage module are performed. Progressing one level is highlighted by a successful message vocally announced. Furthermore, the completion of a scheme of difficulty levels in a category receives a reinforcement provided by a short animation in addition to the sound message. The execution of one of the modules can be halted when the user wishes. The next time the user accesses the software, it will resume from the point where it had been interrupted.

## 5.2 Memory

The second module is an electronic version of the classic Memory game: the screen displays a grid containing "covered" cards. The user is invited by a pre-recorded voice to find the matching pair of cards. Next, the user can begin to uncover a pair of cards. When two matching cards are identified, a confirmation tone (trumpet) is played and the two cards disappear. Conversely, if the two cards are different, they are again covered and an audible failure sound is played. At the bottom of the interface there is a progress bar to show the percentage of correct matches performed. At the end of the game, when all the card pairs are found, the whole schema is shown and a success message is vocally announced.

Cards are grouped in categories: objects, animals and shapes. The game starts with the first category, progressing through the others, passing from the simplest scheme (two pairs of objects) to the most complex (six pairs of objects). When the user finishes a schema, a visual and audible feedback is provided, as in the Barrage module.

Each schema is associated with a threshold value, which takes into account the maximum number of attempts to solve a schema. If a student goes over this error threshold, the same pattern is repeated automatically.

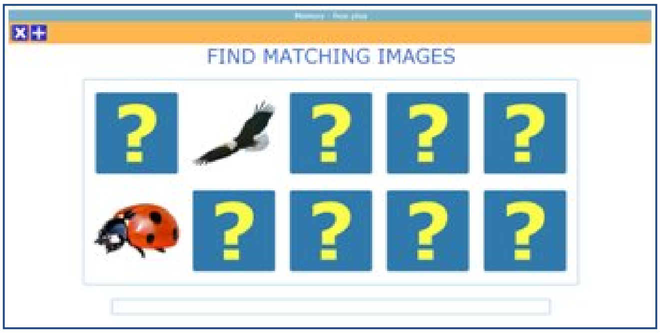


**Fig. 6.** Memory User Interface

### 5.3 Pilot Test

A pilot test has been performed with two adults with DS to refine the software functionalities and improve the user interfaces. In order to be able to generalize our results to wider clinical samples of adults with intellectual disabilities, we included subjects with DS with additional co-morbid diagnoses and/or who are taking prescribed medication. Exclusion criteria were a diagnosis of autism and severe motor and sensory problems, as these were considered to affect pre- or post- assessment (and hence reliability of assessment) or training ability.

The software was described to the participants showing the demo and all the functions. Then each of the two subjects, G. and O., performed the assigned Barrage and Memory modules using a laptop. Each subject executed the modules separately under the supervision of the tutor and the STELLA DS engineer.

Two subjects with different abilities were chosen, in order to cover more functioning levels. Results were quite different and reflected their abilities: one subject executed the trials autonomously; the other performed very slowly, requiring many tutor prompts.

The pilot test highlighted very positive features of the application, and the STELLA DS application was enthusiastically accepted by the testers. Positive points are the attractive images and the vocal feedback: the application "speaks" to the users. One of the users was charmed by the new words, and he repeated them to himself after they were announced. Both students appreciated the vocal reinforcement during the execution of the exercises. Some critical points emerging from the pilot test:

- In some points the demos were not effective and clear as we supposed. We are planning to substitute them with short videos showing the correct interaction with the game.
- During the execution of the Barrage module, images used as stimulus (to indicate the targets) can be confused with the images to be searched. These prompts are important since they offer a visual model when the word is unfamiliar. To clarify the exercise, we have modified the UI, miniaturizing these images, removing their surrounding boxes and creating a visual separation between the discriminative stimulus and the area where user interaction is required; see Fig. 7.


**Fig. 7.** Barrage UI: the revised interface

Currently the application allows tuning the game's level of difficulty based on student responses. Due to the large variability of users in terms of aptitude and skills, it would be appropriate to plan to diversify the trials depending on user abilities. For example, particularly able subjects may avoid too-simple levels of difficulty, which bore them.

## 6 Discussion

Preliminary results of a randomized controlled trial ongoing at IRCCS Stella Maris assessed the effect of an intervention of EE (psycho-physical training regarding functional and cognitive skills) in adults with DS. The good success of previous training experience realized in IRCCS Stella Maris Institute suggests enlarging the number of the trainable people with STELLA DS. Tele-rehabilitation offers cost-effective services that can improve access to care for patients with cognitive disorders.

Tele-rehabilitation through a Web application can be utilized at home to create an Enriched Environment to train the brain and prevent dementia in subjects with DS. Specific rehabilitative content (memory, attention, cognition, reasoning, visual-constructive and spatial abilities) will be trained and the effects of treatment will be tested by a neuropsychological protocol before and after training. We speculate that our software will support the rehabilitation of cognitive function at home, at low cost and with the possibility of increasing the number of involved people with the collaboration of family associations.

The STELLA DS arose from a collaboration of the Institute of Informatics and Telematics of CNR in Pisa and the Stella Maris, a National Biomedical Research Institute for neurological and psychiatric disorders in children and adolescents. The aims of the two groups are dissemination of the importance of a tele-rehabilitative training to a large community of disabled people.

## 7 Future Work

This paper describes the design and development of software for combating the mental decline of people with DS. The software is a web application usable anytime, anyplace, with any device supporting a web browser. The application is partly customizable and is currently available in Italian and English. Two training exercises have been implemented to verify the feasibility of the proposed approach. A pilot test with two users has shown the interest of participants in using the software, confirming the opportunity to investigate the efficacy of the tool for tele-rehabilitation.

For future studies we will develop more training games, and will carry out a user test recruiting a large sample of participants in order to investigate long-term effects of the use of the software and to evaluate its relevance in preserving cognitive ability and individual autonomy in everyday functions.

The effects of cognitive training via STELLA DS will be tested following a neuropsychological protocol that will evaluate the enrolled subject's abilities before and after the training.

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