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The aim of SD Revista Médica Internacional sobre el Síndrome de Down (International Medical Journal on Down’s Syndrome) is, on the one hand, to gather current knowledge on the medical aspects of Down’s Syndrome, and to continuously review and update this, from the most promising advances in basic sciences, such as molecular biology and genetics, to daily clinical practice; and on the other hand, to look at those psychopedagogical aspects, that, due to their relationship with the medical field, could be of practical interest for general and specialist paediatricians associated with Down’s Syndrome. SD will consider publishing clinical or research articles associated with all branches of Down’s Syndrome.
EDITORIAL

A human rights issue

K. Trias Trueta

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When a financial crisis arrives accompanied by a crisis of values as important as the one we are living in today, there are many questions we must ask in order to optimise available resources.

The demands of society make the priorities clear. However, the cut-backs affect the most basic needs of the civil society and, it is precisely the civil society that is “leading” it, as the Universal Declaration of Human Rights proclaims, the defence of the respect for the inherent dignity of all members of the human family and the equal and inalienable rights for all that constitute the foundation of freedom, justice and peace in the world.

To raise awareness of the rights of people with disabilities has been a constant concern of the Foundation. From the beginning we realised that, to create the conditions so that people with intellectual disabilities should be in, and live in, a situation of real equality like the rest of the population, where their opinion counts, was a question of human rights.

The Commissioner for Human Rights, Thomas Hammerberg, highlighted that people with intellectual disabilities should not be deprived of their individual rights. Thus they must be given more support so that they can exercise their rights, and other types of support that gives them equal opportunities to create their life projects.

We have opened a series of training and mutual skills spaces on human rights for young people and adults who increasingly feel like citizens in their own right. Because to speak about human rights simply means accompanying and helping to reflect on our role in society. To help put our particular situation in the world into context, to be aware of other realities –difficult and complicated– that also co-exist with us. A diverse society requires that its members are educated in human rights in order to help build a better society, based on fraternity, solidarity and equality.

Knowledge provides us with the means to develop a more coherent discourse on our needs and, thus, to give more confidence to the parents, caregivers, guardians or therapists due their yearnings for independence. The interest of the individual must be taken into account and to make the most of their level of maturity and understanding, whatever it is. And when the skills of the individual are not sufficient –due to not being able to assess danger and the protection of their safety–, their rights will have to be defended by their representatives, but always trying to involve the individual to take part. To work for individuality of the person involves helping them understand better so that they can grow with personal security and develop their own opinion that will enable them to make decisions to achieve their life projects. Hence the importance of a service like that of an Independent Life offered by the Foundation, in which its principles are based on making these projects a reality, and where the secret is the support that, besides being individualised and respectful to the person, must be based on accompanying to ensure that it is the centre of all planning.

Because to accompany is not to walk instead of the other, nor to advance it, however good is the intention. To accompany is to support, to know how to act so that the individual advances and progresses. It is not to teach how to live, but to help to live. We all learn from one another.

We have self-managing groups in the Foundation that meet to debate and defend their rights. We have created the EXIT21 blog as a platform to give them a voice and to announce their decisions.
Hypothyroidism and Down’s syndrome

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KEYWORDS
Hypothyroidism; Down’s syndrome; Thyroids; Infancy; Subclinical

Abstract
Introduction: Subclinical hypothyroidism is common in the first years of life of children with Down’s syndrome (DS). The aim of this study was to analyse the natural evolution of this disease and to identify the factors that predict its spontaneous remission.

Material and methods: A retrospective, observational study conducted on patients with DS and hypothyroidism diagnosed before 5 years of age, who were seen in a DS reference medical centre.

Results: A total of 53 patients, 28 boys and 25 girls, with a mean age 2.4 ± 1.1 years, were identified with subclinical hypothyroidism. The hypothyroidism resolve spontaneously in 39 cases (73.6%), in a mean time of 13.2 ± 11.1 months, this resolution rate being significantly higher in the patients without goitre: 94.9% (95% confidence interval [CI]: 81.2-99.3%) vs 28.6% (95% CI: 4.4-37.7%), p < .05, and with negative antithyroid antibodies: 89.7% (95% CI: 74.6-96.2%), vs 42.9% (95% CI: 20.7-56%), p < .05).

Fifteen patients (28.3%) were treated with levothyroxine.

Conclusions: The subclinical hypothyroidism that appears in early infancy in DS is usually transient. The absence of goitre and antibodies is associated with a higher spontaneous resolution rate.
Introduction

The prevalence of medical disorders in individuals with Down’s syndrome (DS) is higher than in the general population, and has a negative impact on their quality of life and life expectancy. Thyroid disease features prominently among these medical problems. Both hyper- and hypothyroidism, mainly of autoimmune origin, occur more often in DS1-3, the latter being 6 times more common than the former4,5.

As well as the increased risk of developing hypothyroidism with age2-6, children with DS have a higher probability of presenting with two other thyroid problems in their first months of life: congenital hypothyroidism7, which can be easily detected by neonatal screening, and, more frequently, an isolated mild increase in thyrotropin (TSH) or subclinical hypothyroidism8,9). This latter disorder is usually transient and rarely progresses to clinical stages, and normally self-resolves without the need for treatment10,11. But there are few systematic studies on this disorder, and the factors associated with this self-resolution are not clear.

There are also doubts on the possible benefits of hormonal treatment on the development of these patients, despite it being a subclinical, and even transient, disorder. In this sense, the results of a double-blind randomised clinical trial with 196 children with DS were innovative compared to previous studies. Based on the theory that all subjects with DS are slightly hypothyroid at birth12,13, this study evaluated the effect of systematic treatment with levothyroxine started in the neonatal period and during the first two years of life, compared with placebo. The treatment with levothyroxine showed a slight improvement in psychomotor development and somatic growth at 24 months, and thus concluded that hormonal treatment should be considered in neonates with DS to obtain optimal development and growth14.

The primary aim of the current study is to characterise and analyse the progression of the hypothyroidism diagnosed in a population of children under 5 years-old with DS, and to determine the main factors that predict the spontaneous resolution of the disorder.

Material and methods

Patients

A retrospective study was conducted on a clinical series of patients with DS and hypothyroidism. The data were collected by reviewing the clinical histories of patients with DS from the Fundació Catalana Síndrome de Down (FCSD), registered between 1993 and 2008. The FCSD, established in 1984, is a non-profit organisation that has as its aim to promote the full development of subjects with DS in Catalonia. It provides support to patients from prenatal diagnosis to adulthood, but does not have a complete populational record of all patients with DS. The medical program of the Foundation includes the annual systematic screening for thyroid diseases, including the annual determination of TSH. The FCSD data base was analysed, and all patients with hypothyroidism were identified, whether it was clinical (elevated TSH with decreased levels of total triiodothyronine (TT3) and/or free thyroxine (FT4), or subclinical (elevated TSH with normal TT3 and FT4 levels). All patients were less than 5 years-old at the time of diagnosis, and had been assessed by the same endocrinologist.

Clinical data

Among the variables collected from the clinical histories included: age when diagnosed with hypothyroidism, sex, the presence of thyroid disease in the family history, the...
Results

Baseline characteristics

Out of a total of 1903 clinical histories from the FCSD, 149 patients were identified with an alteration in thyroid function, which is a prevalence of 7.8% (95% CI: 6.6-9%), 12 of them with hyperthyroidism, and 137 with hypothyroidism (fig. 1). Of the 137 patients with hypothyroidism, 54 (28 boys and 25 girls) were diagnosed before 5 years of age; 1 of them had clinical hypothyroidism with a low FT4, while the remaining 53 fulfilled the criteria of subclinical hypothyroidism, and were included in the study. The follow-up time was 54 ± 19 months. The mean age was 2.4 ± 1.1 years. Table 1 shows the baseline characteristics at the time of diagnosis.

There was a family history of thyroid disease in 13 patients (24.5% [95% CI: 12.3-37.7%]), multinodular goitre and hypothyroidism in the majority of the cases, with a predominance of the maternal line. Karyotype analysis was available in all cases, which confirmed trisomy 21 as the most common genetic abnormality (n = 48, 90.5% [95% CI: 80.5-97.3%]), followed by mosaicism (n = 4, 7.5% [95% CI: 0.4-14.4%]) and 14/21 translocation (n = 2, 3.7% [95% CI: 1.3-8.7%]). There was a high prevalence of concomitant medical disease, particularly congenital heart diseases, which affected 23 cases (43.4% [95% CI: 29-56.2%]).

Signs and symptoms

A total of 19 patients (35.8% [95% CI: 22.4-47.9%]) had some symptom or sign of hypothyroidism on diagnosis. The most frequent symptoms described were, constipation (23.6%), rough, dry or cold skin (22.1%), and weight increase (11.1%). Other, less frequent, symptoms were: intolerance to cold, decrease in sweating, asthenia, hearing loss, or drowsiness. The mean score on the Billewicz scale was −13.9 ± 11.9 (non-specific). There were no differences in the mean values of TSH between the patients with and without symptoms (mean 8.2 ± 2.9 μU/mL vs 8 ± 2.8 μU/mL, respectively, p = .5). The neck examination showed the presence of goitre in 12 cases (22.6% [95% CI: 15.2-65%]), which, in the majority of cases (91.7%), was of low grade (Grade I).

Anthropometric data

The mean weight percentile compared to the Spanish population with DS was 55 ± 24.2 at the time of diagnosis, and a mean length/height percentile of 45.7 ± 26.6. The BMI at diagnosis, excluding patients less than 2 years-old, was 16.5 ± 1.3, with a mean percentile BMI compared to the general
Hypothyroidism and Down’s syndrome

The percentage of overweight patients at the time of the diagnosis was 22.9% (95% CI: 9.1-37%), and there was 11.4% with obesity (95% CI: 0.6-21.4%).

There were no significant differences between the 2 groups in the initial weight and length/height percentiles, nor at one year or two years of onset.

Laboratory data and rate of resolution

The mean TSH at diagnosis was 8 ± 2.8 µU/ml. The subclinical hypothyroidism was resolved in 39 of the 53 cases (73.6% [95% CI: 61.7-85.4%]), spontaneously in 35 patients, and after withdrawing the levothyroxine medication in 4 cases. The mean time required for the resolution was 13.2 ± 11.1 months from the diagnosis. The majority of these cases were resolved between 4 and 5 years (fig. 2). No patient progressed to clinical hypothyroidism during the observation period. In the group in which the hypothyroidism resolved, the mean TSH in the last recorded visit was 3.6 ± 1.9 µU/ml, and the age was 6.8 ± 1.4 years.

Twelve cases (22.6% [95% CI: 11.4-33.9%]), had positive anti-TPO and/or anti-Tg antibodies at diagnosis or during the follow-up.

The factors significantly associated to the resolution of the hypothyroidism were the absence of goitre and antithyroid antibodies at diagnosis and during follow-up. Other factors analysed, but had no significant association were,
age, sex, family history, absence of symptoms, and the mean TSH at diagnosis (table 2). When goitre was absent, the resolution rate was 94.9% (95% CI: 81.2-99.3%), while if it was present, the hypothyroidism resolved in 28.6% of the patients (95% CI: 4.4-37.7%) ($p < .05$). The resolution rate was 89.7% (95% IC: 74.6-96.2%) in the group with negative antithyroid antibodies, while it was 42.9% (95% CI: 20.7-56%) ($p < .05$), in the group with positive antithyroid antibodies.

**Treatment**

A total of 15 (28.3%) patients received treatment with levothyroxine. In the majority of cases, the reason that indicated treatment was a TSH higher than 10 µU/mL (in 1 case there was a decrease in peripheral hormones, and 1 case required cardiac surgery due to a congenital heart defect. The mean age at the start of treatment was 4.4 ± 3 years, and the mean TSH was 10.9 ± 1.3 µU/ml. The mean initial dose of levothyroxine per kilogram was 1 ± 0.4 µg/kg/day (1.1 ± 0.3 µg/kg in boys and 0.9 ± 0.5 µg/kg in girls).

**Discussion**

In the present study it has been observed that the hypothyroidism characteristic of early infancy in DS usually presents as a subclinical disorder. The distribution of the disorder in this initial stage is similar between sexes, which contrasts with that which occurs in the population without DS, where the hypothyroidism is clearly predominant in the female sex$^{17-20}$. The majority of cases are resolved without treatment, and the persistence or progression to clinical stages appears to be linked to the presence of autoimmune factors.

The cause of subclinical hypothyroidism in the first years of life of DS patients is not clear. Among the different hypotheses are that it is due to a local peripheral defect in the production or action of T3$^9$, thyroid insensitivity to TSH$^{10,11}$, or an inadequate secretion, or less active TSH at central level, although in some studies the bioactivity of the TSH molecule appears to be normal in these patients$^{21,22}$. The findings by Karlsson indicated that autoimmunity did not seem to play an essential role in this disorder, although this normally appears later on, at school age$^{17}$. Van Trotsenburg suggested that virtually all individuals with DS had a congenital defect in the regulation at the level of the thyroid gland itself, which would be in a direct relationship with the trisomy state of chromosome 21. This approach is the result of a longitudinal study conducted on 97 neonates with DS, who were followed-up until up to 26 months of age. The results showed that the distribution of the TSH and FT4 values were normal or Gaussian, but dis-

**Table 2** Factors associated with the remission of the subclinical hypothyroidism

<table>
<thead>
<tr>
<th></th>
<th>Hypothyroidism remission</th>
<th>Hypothyroidism persistence</th>
<th>$p$</th>
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</thead>
<tbody>
<tr>
<td>n (%)</td>
<td>39 (73.6%)</td>
<td>14 (26.4%)</td>
<td>–</td>
</tr>
<tr>
<td>Mean age at diagnosis (years)</td>
<td>2,5 ± 1,1</td>
<td>2,3 ± 1,2</td>
<td>0,95</td>
</tr>
<tr>
<td>Mean age at the last recorded visit (years)</td>
<td>6,8 ± 1,4</td>
<td>6,6 ± 1,4</td>
<td>0,67</td>
</tr>
<tr>
<td>Sex (% girls)</td>
<td>47,4%</td>
<td>50%</td>
<td>0,29</td>
</tr>
<tr>
<td>Family history of thyroid disease n (%)</td>
<td>8 (20,5%)</td>
<td>5 (35,7%)</td>
<td>0,33</td>
</tr>
<tr>
<td>Hypothyroidism symptoms/signs n (%)</td>
<td>11 (28,2%)</td>
<td>8 (28,6%)</td>
<td>0,72</td>
</tr>
<tr>
<td>Mean TSH at diagnosis (µU/ml)</td>
<td>3,6 ± 1,9</td>
<td>9,1 ± 3,5</td>
<td>0,13</td>
</tr>
<tr>
<td>Absence of goitre n (%)</td>
<td>37 (94,9%)</td>
<td>4 (28,6%)</td>
<td>&lt; 0,05</td>
</tr>
<tr>
<td>Negative TPO/Tg antibodies n (%)</td>
<td>35 (89,7%)</td>
<td>6 (42,9%)</td>
<td>&lt; 0,05</td>
</tr>
</tbody>
</table>

$Tg$: anti-thyroglobulin; $TPO$: thyroid peroxidase antibody; $TSH$: thyrotropin.
placed to the right and to the left, respectively\textsuperscript{11}. Along the same lines, another recently published study also showed that the TSH levels in neonates with DS were higher than those in the control group, particularly in the males\textsuperscript{21}.

In these cases of subclinical hypothyroidism detected in these initial stages, the most usual approach consisted of performing an analytical follow-up with no initial therapeutic intervention, given its frequent remission\textsuperscript{16}. The longitudinal study by Gibson showed that only one of the 20 cases of children with DS and an isolated increase in TSH developed clinical hypothyroidism in a second hormone determination performed 4-6 years later. Furthermore, the TSH returned to normal in the majority of patients in a short period of time\textsuperscript{16}. Similarly, in our study only 1 patient had decreased T3 and T4, and the disorder resolved spontaneously in the majority of cases.

The presence of goitre or antibodies suggests the development of an autoimmune thyroiditis process, of such a diffusely in the majority of cases. Decreased T3 and T4, and the disorder resolved spontaneously in the majority of cases.

In conclusion, hypothyroidism in the first years of life in patients with DS is shown as a subclinical and mainly spontaneous disorder, with the principal predictive factors of spontaneous remission being the absence of goitre and anti-thyroid antibodies.

\section*{Acknowledgements}
To Reyes Alcoverro, Secretary of the FCSD, on helping us with the task of collecting data for the study.

\section*{Conflict of interests}
Authors declare not to have any conflict of interests.

\section*{References}


CASE REPORT

Down’s syndrome with abnormal inner ear: Is it suitable for cochlear implantation?

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KEYWORDS
Down’s syndrome; Hearing loss; Cochlear implant; Large vestibular aqueduct syndrome

Abstract
Hearing loss is a common problem in Down’s syndrome (DS). The majority of this population, up to 80%, are suffering from a conductive type hearing loss, whereas estimating 4-20% are due to sensorineural hearing loss. Over the years, the treatment of profound sensorineural hearing loss has been changed since the introduction of cochlear implants. We report a case of a 4 years and 5 months old child with DS and low Intelligence Quotient that had been referred to our centre for cochlear implants. In view of late referral and multiple additional handicaps, with addition of having Larged Vestibular Aqueduct Syndrome (LVAS), bilateral incomplete partition of cochlear Type II and abnormal periventricular white matter, she had been rejected for cochlear implantation.

PALABRAS CLAVE
Síndrome de Down; Hipoacusia; Implante coclear; Síndrome del acueducto vestibular dilatado

Síndrome de Down con oído interno anómalo: ¿es apto para un implante coclear?

Resumen
La hipoacusia es un problema frecuente en el síndrome de Down (SD). La mayoría de esta población, hasta un 80%, sufre hipoacusia conductiva, mientras que el 4-20%, según las estimaciones, corresponde a hipoacusia neurosensorial. A lo largo de los años, el tratamiento de la hipoacusia neurosensorial profunda ha cambiado desde la introducción de los implantes cocleares. Presentamos el caso de una niña de 4 años y 5 meses de edad con SD y un bajo cociente intelectual, que fue remitida a nuestro centro para ser sometida a implantes cocleares. En vista de la derivación tardía y las múltiples discapacidades adicionales, además de la presencia de síndrome del acueducto vestibular dilatado (SAVD), partición incompleta bilateral coclear de tipo II y sustancia blanca periventricular anómala, no se consideró adecuado el implante coclear.
Introduction

Down’s syndrome (DS) is the most common genetic disorder, occurring in approximately 1:800 live births. Children with DS have altered head and neck structure that results in increased otologic, upper airway, and sinonasal disease. Between 38% and 78% of peoples with DS have abnormalities of the external, middle and inner ear have been described, which contribute to the hearing loss in these individuals. Out of this, over 80% of the hearing loss is conductive and this is due to otitis media with effusion, therefore amenable to medical and surgical intervention. However, 4 to 20% of hearing loss in this population is due to sensorineural hearing loss. It was initially thought that individuals with additional disabilities and learning disabilities were not suitable candidates for implantation, but with a growing body of knowledge and good results, inclusion criteria are expanding and increasing numbers of such candidates have been implanted. Many of these individuals, especially those implanted at a young age, do remarkably well due to preservation of the spiral ganglion and successful post operative habilitation.

Clinical presentation

A child with DS and global developmental delay was referred to our centre at 4 years and 5 months of age for audiological assessment as a potential candidate for cochlear implant. The child was diagnosed to have prelingual bilateral hearing loss at 4 months of age and bilateral middle ear effusion. Auditory brainstem recordings confirmed a profound sensorineural hearing loss on the right and moderate to severe hearing loss on the left ear when she was 5 months of age. However, the myringotomy with ventilation tube insertion was performed only at the age of 1 year 4 months old and postoperatively, she has been fitted with hearing aid binaurally. However the usage of hearing aid was inconsistent until 4 years old. At the age of 2 years, serial of re-programming and optimization of hearing aid were performed, however the result of aided response evaluation showed the hearing aid was under amplification. She had a trial of consistent hearing aid for about 5 months, however there was no benefit. She was then referred for consideration of cochlear implant.

High Resolution Computed Tomography (HRCT) imaging of the temporal bone performed at 4 years and 9 months of age revealed a large vestibular aqueducts bilaterally (fig. 1A) and bilateral incomplete partition of cochlear Type II (fig. 2). There was also fluid within mastoid air cells, both middle ears and both epitympanic spaces.

The magnetic resonance imaging (MRI) demonstrated an enlarged endolymphatic sacs bilaterally (fig. 1B) with normal 7th and 8th nerves, internal auditory canal, vestibules and semicircular canal. There are multiple dilated periventricular region in both temporal and parietal lobes likely represents incomplete myelination and steep straight sinus with absent sagittal suture (not well demonstrated in MRI) suggestive of brachicephaly (fig. 3).

Discussion

Cochlear implantation is the treatment frequency of severe to profound sensorineural hearing loss. More candidates had been implanted at younger age with good capacity to develop language at a rate equal to that of their hearing peers. Previously due to limited studies on outcomes for this implantation procedure, the candidacy criteria were very strict. However, recent serial studies have shown good outcome from this invasive procedure, and the indications for implantation have gradually been revised. Now there are more implant devices licensed for use in children as young as 12 months and in additional and learning disabilities. The children with additional disabilities can potentially broaden their communication skills, and make progress, though possibly at a slower pace than children without additional disabilities.

Patient with DS and hearing loss posed a major challenge to the successful use of hearing aids and other rehabilitative devices including cochlear implants. They can have multiple additional handicaps, including learning and communication dif-

![Figure 1](image1.png)  
**Figure 1** Axial high resolution computed tomography temporal bone (A) showing dilated vestibular aqueduct on both sides (arrows) and axial magnetic resonance imaging T2WI (B) demonstrating enlarged endolymphatic sac bilaterally (arrows).
Down's syndrome with abnormal inner ear: Is it suitable for cochlear implantation?

This group of children have been shown to have an effect on subsequent language development and performance post-implantation, with outcomes below those of implanted children without additional disabilities. A recent survey of The Cochlear Implant Programmes in DS by the British Cochlear Group (BCIG) in 2010, four with DS children have received implants. They reported that all children remain implant users 12 months to 4 years post-implantation with a significant improvement seen as early as from 9 months post-implantation in terms of communication and behavioural outcomes.

This case reported a DS's child with global developmental delay and pre-lingual congenital sensorineural hearing loss. She initially had inconsistent use of her hearing aid. By 20 months, the equipment was being worn more consistently with optimal fitted hearing aid. However the child was not having benefit from the hearing aid, therefore she was referred for cochlear implantation.

Several factors were identified which cochlear implant was not an appropriate intervention for this child. She had later referred to our centre for cochlear implant (at 4 years 5 months old), in which ideal age for referral as early as 3 months old. Susan Willey et al. in 2009 reported that the possible factors of delayed in referral were multi-disciplinary process when deciding whether a child should be referred for an implant, such as degree of hearing loss, marital status of parents, type of insurance, and living in area where income is below the average. In addition to that, an audiologist's ability to determine possible audiologic candidates for referral and managing otolaryngologist who focused on otology were more likely to be referred early compared to children managed by an otolaryngologist who had a wider range of interests.

The other concern about the child is having abnormal inner ear structures and abnormal brain parenchyma. She was rejected for cochlear implantation as HRCT showed bilateral incomplete partition of cochlear Type II. She also has other otological abnormalities, which is LVAS. She is at risk of perilymph gusher intra-operatively and at risk of meningitis post-operatively. However, few reports of several studies have showed benefit with speech recognition to varying degrees from implantation in patients with LVAS and can be offered as an eventual treatment for hearing loss in these patients. In addition, Asma et al. in recent series in 2010, had advocate this group of child should be implanted earlier after discussing pros and cons with parents as they found out duration of profound hearing loss and residual hearing appear to be critical factors in determining implants success. Furthermore, she was suffering from otitis media with effusion (OME), this raise the issue in the candidacy of cochlear implant. Schwartz & Schwartz in 1978 in their study of 38 children's (mean age, 3.1 years) with DS, reported that more than 60% of the series demonstrated otoscopic and acoustic impedance evidence of middle ear effusion. It is postulated that the OME is secondary to atypical head and neck anatomy, including macroglossia, hypoplastic nasal bones, oropharynx and nasopharynx that are narrower volume. In addition, Eustachian tubes are smaller in diameter and at a less acute angle to the hard palate.

There was concern that implantation in the situation of the otitis media prone ear would lead to increased rates of complications, particularly the risk of infection spreading from the middle ear intracranially through the channel created by the cochlear implant. However, Hans et al. and BCIG in their survey in 2010 reported all their patients had OME, no intra-operative or post-operative surgical complications were encountered.

A part from otological abnormalities, she also has global developmental delay and her MRI showed multiple dilated periventricular region in both temporal and parietal lobes likely represents incomplete myelination and features suggestive of brachicephaly. With all the reasons discussed earlier, she has very limited benefit of cochlear implantation, and University Kebangsaan Malaysia cochlear implant committee decided to reject her from the programme. She will learn later for sign language.

As conclusion, DS babies with hearing loss should be encouraged to have consistent audiological followed up and having hearing aid intervention. We would encourage clinicians caring for these children and their families to consider referral for assessment by a Cochlear Implant Programme as early as 6 months of age.
References

CLINICS AND PRACTICE

Skills and social interaction of children with Down’s syndrome in regular education

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KEYWORDS
Down’s syndrome; Social Interaction; Child; Social skills

Abstract
This study identifies the process of social interaction of children with Down’s syndrome (DS) in the regular educational system of a city in the interior of São Paulo, Brazil. Six children aged from three to six years old participated in the study. Each child was videotaped in four situations of social interaction in two distinct environments (indoors and outdoors), which enabled the analysis of interpersonal and self-expression skills through the observation of 15 types of behaviors. The results reveal that the behavior type “Interacts with another child”, within the category “interpersonal skills”, was the most frequent both indoors and outdoors with an average of 27.5 and 28.3, respectively. With regard to “selfexpression skills”, only the behavior “Smiles” had a considerable number of occurrences indoors with an average of 8.16, while the behaviors “Smiles” and “Imitates other children” presented significant occurrence outdoors with averages of 5.16 and 3, respectively. The conclusion is that including children with DS in the regular educational system promotes new forms of learning and interaction for them through daily contact with children with typical development, enabling them to acquire social interaction skills.

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Introduction

Down’s syndrome (DS), or trisomy 21, is a very frequent chromosomal abnormality that occurs due to an extra genetic load from the time of development within the uterus, marking the child for her/his entire life. Confirming trisomy 21 has no prognostic value, since there is a consensus among the scientific community that there are no degrees of DS; developmental differences are due to individual characteristics (genetic inheritance), stimulation, education, environment, and presence or absence of clinical disorders, among others. The mental development and intellectual skills of children with DS encompass a large range between mental retardation and intelligence close to standards considered normal.

With regard to developmental milestones, children with DS experience some delay in comparison to children with typical development, while there is a great variability concerning the speed with which skills are acquired among children with DS. Various factors may cause this delay but the main factor is a lack of early and frequent stimulation in the environment where the child lives. Therefore, living in a demanding environment that promotes a diversity of stimuli and different possibilities for discovery enables individuals’ cerebral reorganization and plasticity. Therefore, the school environment becomes an important space for promoting learning and the development of social skills.

The school, as well as the family, is one of the primary microsystems promoting the development of individuals. It is the second most important social institution for children, especially for those with impairments, because the school is a privileged space for encouraging cognitive functions and social skills.

Social skills are socially acceptable learned behaviors that permit the individual to effectively interact with another and avoid or escape from non-acceptable behavior that results in negative social interactions. An appropriate repertory of social skills or different classes of social behaviors is required for children to satisfactorily interact socially with peers and teachers and deal appropriately with the demands of interpersonal situations. A deficit in the comprehension or performance of the demands of a situation and culture interferes in the quality of social relations. The importance of acquiring social skills is acknowledged by all theories of development, being essential in social adjustment processes both for children with typical development and for those with some disorder/impairment.

Social interactions are seen as educative interactions to the extent they present conditions for individuals to acquire concepts, skills and cognitive strategies that affect social development and learning. Social skills in children and adolescents facilitate the initiation and maintenance of positive social relations, contribute to their acceptance by peers and result in a satisfactory school adjustment.

In a school environment, children with DS aged between three and six years old do not present significant differences in the process of social interactions when compared to children with typical development. There is however a deficit in the assertive social skills of children with DS; they tend to have less initiative, showing more passive social behavior.

This study identifies the social skills of children with DS through the process of interaction with children with typical development and verifies whether behaviors differ given the school environment (indoors and outdoors).

Methods

This is a quantitative non-experimental, cross-sectional study with an analytical descriptive approach and, given its characteristics, can be classified as a multiple case study. All ethical criteria for research involving human subjects were complied with.

Six children with DS aged between three and six years old enrolled in early childhood education programs in municipal schools of the regular education network of Ribeirão Preto,
SP, Brazil participated in the study. There is a small number of participants because the focus is on the quality of these children’s social interactions and also because this is an observational descriptive study.

The children were observed during extracurricular activities (outdoor and large areas) and during academic activities (indoors and restricted areas). There were no directed pedagogical actions during extracurricular activities, which took place in the schools’ sports court, sand box, soccer field and playroom. Outdoor areas were similar in all the schools and had large metal play equipment (slide, merry-go-round, tunnel, seesaw, play house), sand toys (bucket, sieve, shovel) and others, such as balls, dolls, saucepans, cars, and construction toys. In these places, children were free to develop their creativity and interact socially during play.

Academic activities took place in more restricted and closed areas composed of tables and chairs with didactic material and toys. Children in these environments generally remained seated in chairs and were gathered at collective tables to develop structured pedagogical activities proposed by the educator. The use of material such as A4 sheets, activity handouts, crayons, colored pencils, ink and brushes, scrap material, Play-Doh, and glue, among others, were frequently observed.

The children’s behavior was recorded with a digital camcorder focused on the participant; interactions between participants and the researcher(s) were avoided during filming. Filming was initiated when the directed activity began according to data collection protocol used by Anhão11. Each child was filmed in two sessions of 15 minutes each on two different days: one session outdoor and one session indoor.

Data collected during filming were analyzed quantitatively. The two sessions were counted together in order to obtain an average of behavior presented by the participants in each of two environments.

A form created by Anhão11 containing a checkbox for each minute of footage (a total of 15 minutes) for each of the behaviors within the categories of interpersonal skills and self-expression skills was used to record data. Each behavior was checked either as present or absent in each minute of each 15 minutes session of filming. Hence, an (X) was placed in the behavior’s checkbox whenever any of the behaviors described below were observed and the minute when it was manifested was also recorded, according to the following categories:

1. Interpersonal skills: “Interacts with another child”; “Interacts with an adult (educator)”; “Interacts with objects (toys, didactic material)”; “Competes with another person for the educator’s attention”; “Fights or hits”; “Presents self-defense”; “Establishes initial contact with another child”; “Plays together with different objects”; and “Plays together with the same type of object”.

2. Self-expression skills: “Cries”; “Smiles”; “Stays alone”; “Sings”; “Imitates other children”; and “Imitates the educator”.

The signed-rank test adjusted for ties was applied to compare the medians of each skill manifested indoors with skills manifested outdoors12.

**Results**

With regard to interpersonal skills, the results show that the behavior “Interacts with another child” is the most frequent behavior both indoors and outdoors, with an average of 27.5 and 28.3, respectively.

The behaviors “Interacts with objects” and “Plays together with the same object” followed with high frequencies, though their averages are inverted in the two environments. The behavior “Interacts with objects” presents an average of 26 indoors while the behavior “Plays together with the same object” presents an average of 22.5 in the frequency of behaviors presented by the studied children indoors. Outdoors, the behavior “Plays together with the same object” presents an average of 21 while the behavior “Interacts with objects” presents an average of 15.5 in the same environment.

Another very frequent behavior in both environments is “Interacts with the educator”, which presents an average of 17.83 indoors and 7.5 outdoors. The behavior “Establishes initial contact with another person” presented an average frequency of 10.5 indoors and 6.6 outdoors.

The behaviors “Competes with another person for the educator’s attention”, “Fights or hits” and “Presents self-defense” were not very frequent in either environment and presented an average below 1 in both cases.

The results concerning the interpersonal skills presented by children indoors and outdoors are presented with greater detail in Figure 1:

Among the behaviors within the category self-expression skills, only the behavior “Smiles” presented a significant frequency with an average of 8.16 in the frequency of behaviors manifested indoors. The behaviors “Smiles”, “Imitates other children” and “Stays alone” were not very fre-
Table 1 Comparing each skill manifested outdoors and indoors (signed-rank test)

<table>
<thead>
<tr>
<th>Skills</th>
<th>Behaviors</th>
<th>Z value</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Interpersonal</td>
<td>Interacts with another child</td>
<td>0.43</td>
<td>0.670</td>
</tr>
<tr>
<td></td>
<td>Interacts with an adult</td>
<td>1.78</td>
<td>0.075</td>
</tr>
<tr>
<td></td>
<td>Interacts with objects</td>
<td>1.98</td>
<td>0.046*</td>
</tr>
<tr>
<td></td>
<td>Competes with another person for the educator’s attention</td>
<td>0.00</td>
<td>1.000</td>
</tr>
<tr>
<td></td>
<td>Fights or hits</td>
<td>1.41</td>
<td>0.159</td>
</tr>
<tr>
<td></td>
<td>Presents self-defense</td>
<td>0.00</td>
<td>1.000</td>
</tr>
<tr>
<td></td>
<td>Establishes initial contact with other children</td>
<td>0.87</td>
<td>0.462</td>
</tr>
<tr>
<td></td>
<td>Plays together but with different objects</td>
<td>*</td>
<td>*</td>
</tr>
<tr>
<td></td>
<td>Plays together with the same type of object</td>
<td>0.53</td>
<td>0.597</td>
</tr>
<tr>
<td>Self-expression</td>
<td>Cries</td>
<td>*</td>
<td>*</td>
</tr>
<tr>
<td></td>
<td>Smiles</td>
<td>1.57</td>
<td>0.116</td>
</tr>
<tr>
<td></td>
<td>Stays alone</td>
<td>1.71</td>
<td>0.087</td>
</tr>
<tr>
<td></td>
<td>Sings</td>
<td>1.00</td>
<td>0.317</td>
</tr>
<tr>
<td></td>
<td>Imitates other children</td>
<td>1.59</td>
<td>0.113</td>
</tr>
<tr>
<td></td>
<td>Imitates the educator</td>
<td>1.00</td>
<td>0.317</td>
</tr>
</tbody>
</table>

*Significant.
*Not possible to calculate.

Figure 2 Frequency of behaviors concerning self-expression skills in each environment.

Frequent outdoors and presented averages 5.16, 3 and 2.16, respectively.

The remaining behaviors presented average frequencies below 1 as shown in Figure 2.

No significant differences were observed between environments when the behavior categories were compared, except for the behavior “Interacts with the educator”, which more frequently occurs indoors. The results are presented in Table 1.

Discussion

Children acquire and improve cognitive, emotional, and social skills that will accompany them during their entire lives in the school’s different environments. Therefore, including students with special needs within the regular educational system is a complex process and requires the involvement and participation of all members within school organizations. This inclusion should be planned in such a way that students with similar ages participate together in all school activities and are gathered in the same class, even if they have different learning objectives with different degrees of complexity. The development of knowledge concerning interactions among students with DS, their classmates and teachers in the regular educational system is both a scientific and a social need, since it supports characterizing and improving school inclusion processes.

Interpersonal skills

The most frequent behaviors within this category of skills included “Interacts with another child”, “Interacts with objects” and “Plays together with the same type of object”. Children with DS very frequently presented the first two behaviors in both environments. Verbal and non-verbal communication, that is, with another individual and interaction with objects, is a basic element of social development, and the ability to manifest them in a coherent and complementary way is essential for a socially competent performance.

When a child begins to enter other microsystems in addition to the family, the quantity and diversity of interlocutors increases, as well as the opportunities to apply and improve his/her social repertory, which influences in a decisive way his/her acquisition and performance of social skills. Considering the relationship with classmates when the child enters school, a greater variability of models and demands is presented for her/him to acquire new social skills.

It is interesting to study the presence of students with deficiencies in a regular educational environment once children adopt the standards of the group for themselves,
as well as the social interactions that occur naturally between students with deficiencies and the remaining students, focusing on another’s role as mediator of one’s interaction within society\(^4\). It is possible to infer that the studied children learn with their peers when included in extra-curricular activities within the school environment, developing and performing social skills during such interaction.

The behaviors “Plays together with the same type of object” and “Plays together but with different objects” are inter-related and refer to playtime and how these objects are used in this context. When the child plays together but with different objects, s/he presents parallel play and is developing basic socialization skills. When the child plays together with the same object, however, s/he presents cooperative play and is exercising and broadening socialization skills\(^5\). This study shows that only the behavior “Plays with the same object” was frequently observed both indoors and outdoors. This situation suggests that children tend to play with the same object or develop play in order to achieve the same objectives, which requires the use of the same object, when they are in the same environment\(^6\). Such a situation indicates more complex play that encourages social interaction skills.

The behaviors “Competes with another person for the educator’s attention”, “Fights or hits” and “Presents self-defense” show how the participants react to situations of conflict. The fact these occurred with low frequency suggests that children with DS were well-adapted to the school environment and interacted well with their classmates.

Studies show that inclusion benefits children with deficiencies in many aspects, such as in relation to speaking skills, social behavior and academic performance\(^7\). Such a fact may be related to the diversity of stimuli provided within the school environment since a demanding environment that promotes different possibilities of discoveries enables the individuals’ cerebral reorganization and plasticity\(^2\).

The behavior “Interacts with the educator” presented very different frequencies depending on the environment: it was very frequent indoors but infrequent outdoors. This finding suggests that children interact more with their peers when outdoors in unstructured play, while educators are more attentive to the children’s actions, helping them with tasks and encouraging them to play when in the classroom (indoors). Accordingly, children also seek out educators to show them their work. It is important to note that through inclusion in school, children start to spend time with their peers and enlarge their universe of social interaction and present behaviors similar to those observed in children with typical development.

The same is observed in relation to the behavior “Establishes initial contact with another person”. This behavior occurred with a relatively low frequency outdoors in comparison to the indoor context. It is possible that children with DS have a greater difficulty initiating social contact as a result of a lack of experience in environments outside the family’s micro system. In more familiar environments of daily living such as the classroom, such skill may develop more easily. These results suggest that children with DS present a deficit in assertive social skills, which are those that depend on initiative, and develop better passive social skills, or those skills in which the environment plays a determinant role\(^8\). Hence, the school environment facilitates social interactions and consequently the development of skills for children to perform well in society.

**Self-expression skills**

Among the self-expression skills, the behavior “Smiles” presented the greatest occurrence in both studied environments, which suggests that children with DS feel well when interacting with their peers in school activities.

Other behaviors, which like “Smiles” express emotion, such as “Cries” and “Sings” very rarely appear. It does not mean that these behaviors do not occur frequently. We suppose that such behaviors were just not very frequent during filming.

The behavior “Stays alone” was not very frequent either, which likely indicates that children with DS have the potential to interact with their peers when they have the opportunity.

For children, attending school means entering a new world in which they have to progressively acquire increasingly complex knowledge, which will be demanded by society and the bases of which are essential to the education of any individual\(^1\). One of the greatest objectives in early childhood education is to enable children to become more autonomous in the classroom, that is, to internalize rules of social life so they can conduct themselves without disturbing the group; social conformity is sine qua non for an individual to be integrated into a group\(^9\).

In relation to the behaviors “Imitates other children” and “Imitates the educator”, the first presented a much more significant frequency both indoors and outdoors. This does not mean that the educator does not play an essential role in the process of social interaction and inclusion, but children this age demand more interaction with other children who also have the same interests. The educator was present all the time, enabling children to develop their social skills\(^1\). These results suggest that children with DS have a greater tendency to imitate other children’s behaviors rather than to seek a “model” among educators (adults). Hence the importance of the school environment in the inclusion process; the school enables children with DS to have greater contact with children the same age with typical development, which does not occur in the protected environment of special education or even in therapeutic settings\(^1\).

This study’s results make clear that the inclusion of children with DS in the regular education network promotes new forms of learning and interaction through daily life with children who have had typical development, leading to the acquisition of skills necessary for them to perform well in society.

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References