# Abstracts

## 10th World Down Syndrome Congress Lifelong living and learning Dublin, Ireland (19th – 22nd august 2009)

The 10th World Down Syndrome Conference was held last August in Dublin, Ireland. Attendance was high, with over 2,000 delegates from around the globe, including an impressively large and active contingent from Australia.

Our own country had scarce representation. Dr. Serés, the medical coordinator of Centre Mèdic Down (CMD), was there on behalf of Fundació Catalana Síndrome de Down.

Like preceding editions, the 10th Congress addressed a variety of social, medical, educational, and other issues. Both the social events and the scientific sessions were very well organized.

Since it is impossible to summarize all the topics that were covered, we have chosen to publish some of the abstracts that we felt to be of the greatest interest from a medical perspective.

*The editors*

### Iron deficiency and Down syndrome

**L. Rastelli, L. Tremblay, M. Pothos, A. Nair**
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Iron deficiency with or without anemia can impair cognition and affect mood and concentration in children. Down syndrome already presents developmental challenges for the child and their family therefore it is crucial to diagnose and treat iron deficiency as soon as possible. To define the incidence of iron deficiency in a cohort of children with Down syndrome less than 18 years of age followed at a regional Down syndrome clinic in Ontario, Canada.

A retrospective review of patients' screening blood work taken from July 2005 to July 2008 at the Children's Hospital of Eastern Ontario was undertaken. These lab results were taken from patients who attended their routine Down syndrome clinic visits. 234 (139 males (59.4%), 95 females (40.5%) results were available for CBC and ferritin. Of these, 3 patients or (1.3%; 2 males, 1 female) had evidence of anemia (hemoglobin ranging from75-93 G/L). 60 patients of the 234 (25.6%; 53 males, 7 females) had low ferritin (5 to 23 micrograms/L) with no anemia. 170 had normal ferritin levels and 1 had a higher than normal level of ferritin. Only 1 (male) had associated Celiac disease as the possible cause of low ferritin. 26.9 % of the children undergoing routine blood screening demonstrated iron deficiency. We recommend that full blood count and serum ferritin be measured as part of the routine blood work starting in infancy in the Down syndrome population.

Early detection and treatment of iron deficiency can help prevent further compromise of cognitive development.

### Does audit & guidelines help improve the medical care & management of children with Down Syndrome?

**S. C. Puri, I. Rawson**
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In 1998 the Down Syndrome Medical Interest Group U.K proposed evidenced based guidelines for basic essential medical surveillance.

Based on these guidelines we audited our services provided to children with Down syndrome. This highlighted inadequacies, subsequently an audit tool & database of children with Down Syndrome was established in 2004.

To determine if recommendations through guidelines & audit improve the quality of service delivered. Audits were carried every two years; the findings were fed back to the service with recommendations. Comparisons have been through re- audit through four years. The results of the audits are summarised as follows:

| | 2004 | 2006 | 2008 |
|---|---|---|---|
| Growth Plotted | 81 % | 94.6 % | 100 % |
| Cardiac Status established | 97.3 % | 97.3 % | 97.4 % |
| Thyroid Function Tested | 85.7 % | 97.3 % | 97.4 % |
| Vision assessment | 58.6 % | 86.1 % | 79.4 % |
| Hearing assessment | 80.6 % | 94.6 % | 100 % |
| Cervical spine instability discussed | 3 % | 43.2 % | 82.4 % |
| Coeliac disease | 3.7 % | 11.1 % | 8.6 % |
| Dental Care discussed | 3 % | 8.3 % | 82.4 % |
| Social Issues discussed | 24 % | 91.2 % | 33 % |

The results demonstrate that guidelines and audit does improve care & service delivery. There has been a successive improvement; however the vision assessments & discussion of social issues have declined. This reflects the withdrawal of the services from within the dedicated clinic due to lack of resources. We recommend a standardised format for follow up and an audit tool based on the guidelines to improve the medical surveillance & management of children with Down syndrome.

## Survey On Risk Factors For Latex Allergy In Children And Adults With Down Syndrome

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Latex allergy can pose a serious health threat children and adults with Down Syndrome (DS). It is a progressive allergy; each exposure to latex increases the sensitization and the risk for developing an anaphylactic reaction. The risk factors that increase the potential for developing a latex allergy are a history of allergies, food allergies, multiple surgical procedures, exposure latex products, and allergic reactions to certain fruits.

A survey was developed to assess risk factors for developing a latex allergy in children and adults with DS. The survey was distributed to parents and caregivers of children and adults with DS in the United States, Canada and the United Kingdom.

325 parents and caregivers of children and adults with DS responded to the survey. The survey findings indicated that children and adults with DS have multiple risk factors for developing latex allergy. In our survey sample, 64% had one or more surgical procedures, 20% had allergy to one or more medications, 22% had one or more food sensitivities, intolerances or allergies,16% had hay fever, 7% had allergies to band aids and/or surgical tape and 8% had other allergies.

Early recognition of the symptoms and preventative measures are the key to stopping the progression of the latex allergy. Early childhood surgery, multiple surgical procedures, allergic reactions and sensitivities are the risk factors for developing a latex allergy. It is imperative raise the awareness of the risk factors for latex allergy in children and adults with DS.

## The Role Of Chlorhexidine In The Management Of Periodontal Health In People With Down Syndrome

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Periodontal disease results in premature tooth loss in some people with Down syndrome.

This study aims to test the impact of the use of chlorhexidine products alongside regular professional cleaning, on periodontal health and patient quality of life.

After ethical approval, patients were recruited as an opportunistic sample from those already attending the Dublin Dental Hospital as well as through Down Syndrome Ireland. Oral and dental health was assessed at baseline and 3-6 monthly until Spring 2009, with the following indices: dental caries-dmft/DMF, Modified Gingival Index, Pocket probing depth, Gingival Bleeding Index, Calculus Index and the Plaque index. Quality of life was assessed using a tool modified by Allison et al. Patients attended 3- or 6-monthly for professional oral prophylaxis, supplemented by either 1% or 40% chlorhexidine varnish, in a double blind cross over design.

29 patients were recruited;3 were lost to follow-up. Data will be presented on the clinical outcomes as well as the quality of life assessment. Based on the study outcomes, recommendations will be made for the optimal frequency of dental visits and the most appropriate adjuncts to supplement toothbrushing. This study was supported by a grant from the Oral and Dental Research Trust, London, UK.

## The Metabolic Syndrome Characteristics In Down Syndrome

**A Tenenbaum, Y Malcah, I Wexler, R Brooks, F Levy-Khademi**
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The metabolic syndrome (MES) and its characteristics obesity, hypertension, dyslipidemia and insulin intolerance are major risk factors of developing cardiovascular diseases, diabetes and Alzheimer's disease. In the general young population its prevalence is 4-5%, and 49% in the severely obese. Dietary modifications with physical activity may lower the prevalence of MES. Down Syndrome (DS) is a possible risk factor for developing MES due to typical medical conditions, e.g. lower basal metabolic rate, hypothyroidism, obesity and insulin resistance.

To determine the prevalence of the MES Characteristics in children with DS visiting a referral center, and to identify possible subgroups with increased risk of developing MES.

99 children visiting the Down syndrome medical center at the Hadassah University medical center in Jerusalem were evaluated for weight, height and blood-pressure, and most of them for fasting glucose, lipids and physical activity as well. The control group included 31 obese patients from pediatric clinics. Overweight children with DS were compared with non-overweight children with DS and non-syndromatic overweight children. 57.6% of children with DS were obese, BMI 85%, 27% had severe obesity BMI 95%.

The overall MES incidence was 8.3% in overweight children with DS compared with 14.3% in non-syndromatic overweight children.

There were no statistically significant differences in MES characteristics between overweight children with DS, non-overweight children with DS and overweight non-syndromatic children.

Testing for MES characteristics in children with DS leads to early diagnosis, followed by adequate treatment which may improve quality of life and longevity.

## An Audit Of Health Service Provision For Children With Down's Syndrome in the UK

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Children diagnosed with Down's syndrome require medical follow-up and screening for a range of associated health problems. This audit looked at how the healthcare needs of these children were managed across the U.K.

To audit how healthcare is provided and whether medical surveillance guidelines are followed for children with Down's syndrome in the U.K.

A web-based questionnaire was distributed to clinicians in U.K. NHS Children's Services via email. (Telephone follow-up was subsequently required due to initial low response rate.) There was a response rate of 34%. Of those who replied, 10% did not know how many children were treated/reviewed in their area, and 21% had no way of ensuring

follow-up of all children with Down's syndrome. Most (86%) included children with Down's syndrome in a general neurodevelopmental clinic, rather than a separate Down's clinic. Over 40% of the children's services had other healthcare professionals present at clinics some of the time. The majority (85%) followed standard guidelines for medical surveillance, and all but two were aware of the Down's Syndrome Medical Interest Group's 'Guidelines for Essential Surveillance'. (20% of respondents were DSMIG members.) 40% of children's services audited their healthcare provision. A significant number of Children's Services in the U.K. had no method of ensuring follow-up of all children with Down's syndrome, which is a cause for concern. However, where follow-up occurred, the majority followed appropriate guidelines for medical surveillance. Recommendations: institute uniform methods of ensuring appropriate follow-up of all children with Down's syndrome; audit parental perceptions of healthcare provision.

## Is Early Onset Of Osteoporosis In Women And Men With Down Syndrome Based On Different Factors? Down Syndrome (DS) Is Associated With Many Biochemical, Metabolic, Immunological And Musculoskeletal Disorders

**M Sustrova**
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The aim of this study was to evaluate bone mineral density (BMD) and find some of risk factors in young adults of DS in both sexes.

We examined 102 persons with DS (50F/52M), mean age W 25, 7±10 yrs, M 27, 8±10 yrs and 40 healthy men. We evaluate the concentration of 25-(OH) vitamin D, 1,25-(OH)2 vitamin D, FSH, DHEA-S and testosterone by Immonotech, BMD by dualenergy X ray absorptiometry.

Concentration of 25-(OH) vitamin D was significantly decreased in most subjects with DS, but not of 1, 25-(OH)2 vitamin D. The concentration of 1,25-(OH)2 vitamin D, were in women with DS significantly decreased as in men ($p<0,001$), also severe and moderate deficit was more often in women. The concentration of DHEA-S in DS men were significantly lower ($p<0,001$) and concentration of FSH were higher ($p<0,001$) that in healthy men, but not of testosterone. Decreased BMD had 60 % of men and 33 % of woman, but BMD and Z score were significantly decreased in men: FN BMD ($p<0, 05$), FN Z score ($p<0, 0001$), L1-4 BMD ($p<0,003$), Z score ($p<0, 01$). Bone mineral density of neck and lumbal spine in men correlated positive with concentrations of DHEA-S and FSH.

90,8% of persons with DS had 25-(OH)vitamin D deficit, in women deficit is severest. In men is low BMD more frequent (60 %) and is caused mainly by hypogenitalism – sexual immaturity and by vitamin D deficit. In both sexes of DS adults is a complex of other factors – changed bone markers and cytokines, health conditions, medication, low physical activity and dietary practices - which may contribute to the early incidence of osteoporosis.

This work was supported by Ministry of Health of Slovakia (No.2005/39-SZU-17)and CENDO.

## Age Related Health Patterns Among People With Down Syndrome In Europe

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Health disparities are evident when people with Intellectual Disability are compared with their peers. While life expectancy among people in this segment of the population is increasing, they are likely to incur age-related physical and mental health risks. To date, people with ID, specifically people with Down Syndrome (DS) have been mostly absent from public health scrutiny in Europe.

To identify age related health patterns in people with DS in 14 countries of Europe who were participants in a wider study that applied a set of health indicators devised to measure aspects of the health of adults with ID.

Information on demographic characteristics,

health status, health determinants was gathered using the POMONA Survey Protocol. Participants were N=158 individuals with DS. Data was collected by means of face to face interviews, telephone surveys and postal surveys.

Data were analyzed according to 3 age-groups: 19-34, 35-54 and 55 years and older. Frequency of sensory and mobility impairments as well as oral health problems, mental health problems, BMI and presence of epilepsy increased with age. Frequency of health checks increased with age while uptake of some checks e.g. testicular examinations was low. Findings suggest that a dedicated set of health indicators has some utility in identifying age-related health patterns among European adults with DS with results suggesting an increase in the frequency of health problems with age. Further developments in building reliable, sustainable health information systems are

needed to inform policy and health service providers.

## Down Syndrome And Immune Abnormalities

**L Tremblay, L Rastelly, M Pothos**
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Previous research in this population has identified several defects in both humeral and cellular immunity. A clinical study of children with Down syndrome in a community setting which documents the correlation between infection rate and severity and known laboratory immunologic abnormalities in this population has not yet been established.

To determine the incidence of immune abnormalities and the correlation with infection rate in a cohort of children with Down syndrome under the age of 18 years followed in a regional Down syndrome clinic in Ontario, Canada.

A prospective study utilizing an inception cohort of children with Down syndrome followed at the Children's Hospital of Eastern Ontario from Jan 2002 to Apr 2003.

Immunologic testing was done once during this time period when the child was infection free for 6 weeks. A weekly symptom diary was completed by the family for one year. Low levels of WBC (34%), lymphocytes (24%) and PMNs (16%) were documented in 64 children sampled. Low levels of CD3 (74%), CD4 (60.4%), CD8 (86%), CD19 (91%), and CD16/56 (53%) were found in 43 children sampled. Correlation with rates of infection from symptom diaries will be described .

Our study confirms a high incidence of immune abnormalities in the pediatric DS population, however, there was no correlation between serious infections and the immune abnormalities described.

## Adults With Down Syndrome Are At Reduced Risk Of Cutaneous Melanoma: Results From A French Study

**D Satge**
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The frequency of cutaneous tumours is not well known in persons with Down syndrome (DS). Review of cutaneous melanoma in DS in France and in the published literature.

This review comprised 3 steps. First, we searched for cases of cutaneous melanoma in the records of 1,417 adults with DS aged 20+years at the Institut Jérôme Lejeune. Disease progression was evaluated. Next, INSERM data (Institut National de la Santé et de la Recherche Médicale) were reviewed to identify melanoma deaths during the period 1979-1999 in persons with DS for comparison with the general population by standardized mortality ratio. Finally, a review of the literature was performed to identify other reported cases of melanoma among persons with DS.

Two subjects at Institut Jérôme Lejeune developed a cutaneous melanoma; a 45-year-old woman with a melanoma of the finger, and a 29-year-old man with melanoma of the foot. The melanomas were removed by surgery and both patients are alive, without progressive disease, at 13 and 3 years, respectively.

Review of the INSERM data indicated 2 observed melanoma deaths where 7.081 were

expected (SMR 28.24; 95%CI: 3.42, 102.03), suggesting a more than 3-fold decreased risk.

In addition, the review of the published literature identified a further 6 reported cases of melanoma in DS, bringing the total to 10 cutaneous melanomas, and no case of non-cutaneous melanoma.

On the basis of this review, persons with DS seem to be at reduced risk of cutaneous melanoma.

## Thyroid Disease In Down's Syndrome Children: TSH Screening In Scotland Using Dried Blood Spot Samples 1997-2007

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Thyroid function surveillance is justified in Down's syndrome (DS) because of the increased incidence of autoimmune thyroid disease. Since 1997 dried bloodspot samples from DS children have been analysed for elevated thyroid stimulating hormone (TSH) levels, indicative of possible thyroid function changes.

Samples are processed in the national newborn screening laboratory. The current method is the AutoDELFIA neonatal hTSH fluoro-immunoassay (cut-off 4mU/L). Results above the cut-off trigger referral to a named paediatrician and clinical and biochemical assessment, including venous thyroid function and autoantibody tests.

In 1997 183 children were screened, with 10 children referred from two Scottish regions. By 2007 the uptake of screening had increased to 602 from 15 different areas. Between 1997 and 2007, 106 children have been referred with TSH elevation at a median age (range) of 10.14 (0.03-17.91) years. Median (range) capillary TSH elevation at referral was 5.69 (4.0-169.0) mU/L. Thyroxine treatment was started in 55% of referrals. Using cumulative data (1997-2002), the prevalence of hypothyroidism in Scottish DS children was calculated as 6.7% with a median annual incidence of 2.4% after the first year of life.

Dried blood spot TSH screening is widely available. Capillary sampling is easily performed, minimising trauma. Acceptability to both children and clinicians, and feasibility is reflected in increasing uptake. Children with mildly raised TSH and normal free T4 can be left untreated while maintaining or increasing surveillance. Annual screening using the newborn screening laboratory is an effective method of detecting developing thyroid disease in DS children.

## The Age Distribution Of Onset Of Celiac Disease

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Celiac disease is being recognized more commonly in the Down syndrome (DS) population with an estimated prevalence of 5%. However, more information is required about the age distribution of onset of celiac disease, natural history and associated comorbities in the DS population.

To determine the prevalence and age distribution of onset of celiac disease in a community based cohort of children with Down syndrome followed in a regional Down Syndrome Clinic at the Children's Hospital of Eastern Ontario (CHEO), Canada. We performed a retrospective chart review of all patients followed at CHEO's DS Clinic from the clinic's inception in 1992 to December 2008. Greater than 97% of the children with Down syndrome living in the CHEO catchment area are followed in this clinic from infancy to age 18 years for their routine DS related medical care. Routine celiac screening is done at ages 2-3 years and repeated subsequently every 2-3 years until age 18 years.

354 patient charts were reviewed. 18 (5%) patients were diagnosed with celiac disease, initially from positive celiac screening, and subsequently confirmed by biopsy. 78% of patients developed celiac disease between ages 6-12 years with negative celiac screening prior to that age. 50% of patients had associated hypothyroidism and/or insulin dependent

diabetes as autoimmune comorbidities prior to the development of celiac disease. Further epidemiologic studies are needed to determine the best timing for screening of celiac disease in the Down syndrome population. DS individuals with associated hypothyroidism and/or IDDM need closer surveillance.

## Alopecia Areata Is A More Frequent Complication In Children with Down Syndrome

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Alopecia areata is a more frequent complication in children with Down syndrome being observed with a higher incidence than in the general population. Some of the resumed etiologies are vitamin A and zinc deficiency and hypothyroidism, but most of the cases are considered to be an autoimmune disorder. 731 patients of our Preventive Medicine Clinic for Children with Down Syndrome (children up to the age of 18 years) were surveyed in regard to the diagnosis of alopecia.

17 (2,3 %) had episodes of alopecia areata. Causative factors could be found in two (sequelae of the treatment of lymphoblastic leukemia and celiac disease, respectively). In 15 patients no apparent etiologic mechanisms could be found. Nevertheless, 13 children were successfully treated (one with celiac disease by a gluten free diet and 12 by homoeopathic treatment). To find specific options of therapy it is important to look for causative mechanisms; for symptomatic treatment homoeopathy is a frequent alternative in our experience.

## Growth In Children With Down's Syndrome And Heart Malformations

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Children with Down's syndrome (DS) have well-documented growth retardation and an increased incidence of heart failure, a known cause of short stature. DS is associated with congenital malformations, with heart malformations being the most frequent in children with DS. Heart malformations are found in approximately 40–60% of DS patients.

To investigate the effect of heart malformations on growth and weight in children with DS. In a retrospective cohort study, data were collected from German children with Down's syndrome by means of a standardized questionnaire. Statistical analyses were carried out using a linear test model. $p<0.05$ was considered statistically significant.

We analysed 7823 growth data sets from 1032 children with Down's syndrome (53.5% boys and 46.5% girls). The frequency of heart malformations in this cohort was 53%. The appearance of heart malformations in the first year of life was found to have a significant impact on growth ($p<0.05$) and weight ($p<0.001$). Between the ages of 2 and 5 years, height was not statistically decreased in children with heart malformations ($p=0.704$) but their body weight was ($p<0.05$). No difference in height was noted for children aged 6 to 10 years with heart malformations ($p=0.58$) but weight remained lower ($p<0.01$).

Children with Down's syndrome and heart malformations show significantly decreased height and weight in the first year of life. After correction of the defect, usually during the first year of life, children show catch-up growth, but body weight remains lower.

## Achondroplasia And Down Syndrome: A Case Report of a Rare Association

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Achondroplasia and Down Syndrome is a rare association of two independent disorders, that has only been reported few times previously. They are distinct genetic diseases, but with overlapping features such as short stature, developmental delay or hypotonia.

To present a clinical case of this rare association to highlight the difficulties found and to help others in the future to deal with these cases. We report the case of an 8 year old girl, born from a mother with achondroplasia and a healthy father. Since achondroplasia is dominantly inherited, the child was expected to have the disorder, but at birth she also had features of Down syndrome as confirmed later by kariotype. We review the evolution of this child both regarding physical health as well as cognitive functions and adaptive behavior during her 8 years of life. We address the problems resulting from the additional burden of having two disorders, and how they can be overcome.

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We report this case not only because the association of these two diseases is unusual, but also to show how the overlapping clinical features may complicate management and follow up.

## Chronic Constipation Due To Dolichosigmoid (Sigma Elongatum) In Children With Down Syndrome

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Chronic constipation is a frequent finding in patients with Down syndrome. It is very often layed aside as a functional disorder specific for people with Down syndrome without considering that there are organic causes which are not only treatable but even may be more frequent than in the general population.

To establish the incidence of dolichosigmoid in children with Down syndrome. We surveyed 731 patients with Down syndrome of our Preventive Medicine Clinic. Beside patients with Hirschsprung´s disease ( 18 ) we were able to make the diagnosis of dolichosigmoid ( sigma elongatum ) in three patients. This disease is considered a treatable congenital or acquired anomaly which can be detected by means of an enema of the colon ( after excluding the diagnoses of Hirschsprung´s disease or other dysganglionoses, respectively).

It has to be emphasized that every case of chronic constipation has to lead to further diagnostic studies to prevent complications and to improve the quality of life of patients with Down syndrome.

## The Influence Of Apolipoprotein E Genotype On The Age Of Onset And Duration Of Dementia In Individuals With Down Syndrome

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Down syndrome (DS) is the most prevalent chromosomal abnormality associated with intellectual disability. The syndrome has features associated with precocious ageing with a high degree of variability among different individuals. Life expectancy of people with DS has improved dramatically, however by 40 years of age virtually all individuals with DS have the neuropathological changes characteristic of an Alzheimer type dementia. Apolipoprotein E (Apo E) is a polymorphic protein which has been associated with the pathogenesis of Alzheimer type dementia with various allelic forms, namely 2, 3 and 4. A negative association has been demonstrated between exhibiting the Apo E4 allelle and the subsequent risk of developing dementia in people with Down syndrome. The purported benefits of possessing an Apo E2 genotype are less clear.

To determine the influence of Apo E genotype on the age of onset and duration of dementia in individuals with DS.

Eighty female subjects with DS and known

Apo E allele status were assessed for the presence of dementia. Those diagnosed with dementia were followed longitudinally, many until the time of death. The age of onset of dementia and subsequent course and duration of the illness were studied with regard to Apo E allele status.

This study presents the assessment findings of those diagnosed with dementia and their decline over time.

## Medical Intervention in People with Down Syndrome -Portuguese Guidelines

**M. Pinto, M. Palha, D. Andrade**
*Associação Portuguesa de Portadores de Trissomia 21 – APPT21, Portugal*

Several countries have developed specific guidelines for the medical follow up of individuals with Down syndrome. However, they must be tailored to the unique characteristics of each country. The Portuguese Down Syndrome Association (APPT21) was responsible for the development of the Portuguese guidelines.

To present the Portuguese guidelines currently recommended by the Portuguese Down Syndrome Association. We made a review of several guidelines from different countries and adjusted the recommendations to our country, taking into account that most people use private services, that we lack support from health visitors and that our population has a lack of information regarding health.

We have ambitious guidelines, trying to cover all medical areas and focusing on the importance of early detections of severe problems. Some aspects are not yet fully settled and are open to discussion. We know that our recommendations have a tight schedule of follow up but that is also true for children without Down syndrome in Portugal. The cost versus benefits of this approach should be discussed. We hope that this presentation will help us to improve even more our medical intervention program in people with Down syndrome.

## Neurodevelopmental Impact Of Congenital Heart Defects In Down Syndrome

**J. Visootsak, L. Huddleston, S. Sherman**
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Nearly half of all children with Down syndrome (DS) are born with a congenital heart defect (CHD). Atrioventricular septal defect (AVSD), the most common form of CHD in DS, occurs in 38-60% of children with DS and CHD, but is observed in only 1 in 10,000 live births without DS. This represents a dramatic 2,000 fold increase in risk for AVSD among individuals with DS compared to those without DS; yet, virtually no studies have examined their neurodevelopmental outcomes.

This study is the first to characterize the early developmental profiles of children with DS and AVSD (DS+AVSD) compared to age-matched children with DS without CHD (DS-CHD).

Participants consist of 2 groups: 6 subjects with DS+AVSD (mean age 15.4 months) and 18 subjects with DS-CHD (mean age 15.6 months). The Bayley Scales of Infant and Toddler Development III was administered by a psychometrician who was blinded to each subject s cardiac status. The Bayley III was administered to the DS+AVSD group after cardiac repair.

The DS+AVSD group exhibited lower composite scores in all domains relative to their age-matched DS-CHD, with the cognitive and language domains showing significant differences, $p < .05$. The motor, social-emotional, and adaptive composite scores were not significantly different between the two groups.

Our preliminary cross-sectional data document that children with DS+AVSD have greater developmental delays especially in the language domain, compared to children with DS-CHD. Implications for outcome and treatment are discussed. Studying young infants with DS+AVSD allows us the opportunity to identify variables linked with early deficits that may lead to further delayed development.

## Is Fatty Acid Intake And Metabolism In Children With Down's Syndrome Different When Compared To Their Non-Affected Siblings?

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Increased incidence and earlier onset of Alzheimer's disease (AD) is well-recognized in Down's syndrome (DS). The reasons for this are poorly understood. In the elderly a diet rich in omega-3 has been shown to reduce the progression of both dementia and AD. Could diet be a significant factor in AD in DS?

To compare fatty acid (FA) intake and metabolism in children with DS, and their nonaffected siblings. Cross sectional study with recruitment via DS Research Foundation and support groups. Inclusion criteria: Confirmed DS, siblings if available used as controls. A 7-day food record was completed for each subject and analysed using Foodbase V4. Blood samples from each child were analysed for FA using thin layer and gas chromatography, and mass spectrometry.