European Down's Syndrome Association

# NewsLetter

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# Editorial

## JEAN-YVES PERCHAT, PRESIDENT

*We are, every day, confronted with our interrogations: how to progress and win the daily battle to build a normal life for our baby, young child, adolescent or adult with Down Syndrome. We often go through hope and despair, but never give up.*

*The reasons why are: solidarity, know-how, love.*

### *Solidarity*

*Our energy is reinforced, amplified, and flooded by the mere existence of the others, i.e.: you, Parents, you, Professionals, you Associations or Societies.*

*Because of the extraordinary capacity of the Medias and the around-the-world-information system, families are discovering other families, other towns, other countries, other cultures, other ways of life, and finding, along the way, we all have the same sense of human being.*

*This is the strengthened expression and the symbol of the unicity of mankind.*

*This is the right way to find in our solidarity the willingness to succeed.*

*This is the beginning of a new time, when the image described at the end of the last century by Langdon Down becomes: "normal", even if: "different".*

*We shouldn't fear anymore the "other's" glance and we should, now, be proud of our child with D.S., as any other of our children.*

*The INTERNATIONAL DOWN SYNDROME CONFERENCE, organised by the American NATIONAL DOWN SYNDROME SOCIETY in conjunction with EDSA, is the first meeting - in the USA - of the Old and the New World.*

*As President of the EUROPEAN DOWN'S SYNDROME ASSOCIATION, I will express the best and warmest greetings from all people with Down Syndrome living in Europe, to all American persons with Down Syndrome, their Families and the concerned Professionals.*

### *Know-How*

*American and European searchers are on the way to discover how the genes are dispatched through the human genome and they probably will tell us, one day - may be at the dawn of the next century -, where they are, how to reach them, and how to correct the "system error" bugging the third chromosome of the twenty first pair.*

*In the meantime, we have to use the best methods and the best programmes available through the world.*

*From that point of view, the fifth International Down Syndrome Conference offers us most recent informations about medical, educational and social know-how concerning people with Down Syndrome. We have to learn from each other and the ability to compare our european methods with the american techniques is a great opportunity.*

### *Love*

*It may be a little bit old-fashioned and out of a normal behaviour to use the word "love" in the context of an international conference, but: could we exist without this non-measurable energy which allows us to help, to stimulate, to improve and find the right answers to the needs of people born with this overdosed chromosome?*

*The power of our action to achieve the potentials of our Down-children has a single motor, and it is the bond who unifies us towards the same direction: We love them.*

NEWS FROM CONGRESSES

# The specificity of Down syndrome

On february 26 and 27, 1993, an International Symposium on "The Specificity of Down Syndrome" was held in Palma de Mallorca, the capital city of the Baleares Islands (Spain). The symposium was organized by the European Down's Syndrome Association (EDSA), the Asociación Sindrome de Down de Baleares (ASNIMO) and the University of the Baleares Islands. President of the symposium was Juan Perera, director of the ASNIMO Centre Principe de Asturias, near Palma de Mallorca.

Many well known speakers gave very interesting lectures on this specific subject.

**Genetic specificity / Isabel Legal**

Dr. Isabel Legal is a researcher at the Human Genome Research Center in Paris, France. She is primarily involved in the genetic mapping of the smallest human chromosome, no. 21, the distal part of the long arm of which causes Down's syndrome. She emphasized that gene expression is proportional to gene dosage. However, that is not always the case. There are examples of non-proportionality too. The cellular membrane protein APP shows a fourfold increase in the cortex upon trisomy of the corresponding gene. This means that regulatory factors can affect gene expression. This might result in a disturbance of normal processes. This is perhaps the case with amyloid plaques, present in patients with Alzheimer's disease.

At the present time it is known that in 95% of cases Down's syndrome is caused by maternal non-disjunction, either in the first or second meiotic division.

One line of research compares rare forms of Down's syndrome, two people with different duplicates, whereupon an attempt is made to link their features to the duplicates. This leads to the so-called physical map. Chromosome 21 contains about 1.5% of the total human genome with 500 - 2000 loci. From these some 30 have been located at the present time. There are five different types of maps made at the moment:

1) the cytogenetic map (by means of colouring techniques the bands and natural break points are made visible); 2) the genetic linkage map (based on family studies); 3) the genetic map; 4) the genotypic-phenotypic map and; 5) the physical map.

When the chromosome 21 project is ready, the same procedures will be applied to the entire human genome.

**Neurons, cerebrum and nervous system / Krystyna Wisniewski**

The second speaker was Krystyna Wisniewski, a paediatric neurologist as well as a pathologist and the Associate Director of Clinical Services and Pediatric Neurology of the Institute for Basic Research of the New York State Office of Mental Retardation. She discussed the consequences of the genetic abnormalities in the structure and function of the nervous system. She stressed the importance of environmental factors in addition to genetic factors. The brain of an individual with Down's syndrome is built somewhat differently anatomically. In addition, its function and biochemistry are somewhat different. Firstly, there is microcephaly: smaller head and lower brain weight. Next, there is the cortical dysgenesis with layers II and IV being particularly different as well as delayed maturation. For adults over 30, there is the effect of the accelerated aging and the increased incidence of Alzheimer's disease.

Dr. Wisniewski explained the phenomenon called apoptosis, which means that 50% of the neurons of a normal brain die before embryonic differentiation is completed. This programmed cell death may be fundamental to proliferation and differentiation of brain cells. But which genes regulate survival? Individuals with Down's syndrome probably have too many "killing genes", genes that are responsible for the mechanism of this apoptosis.

The speaker mentioned one of her own investigations in which the brains of more than 300 boys and girls with Down's syndrome of less than 5 years of age have been measured. She found that 80% of them were microcephalic, whereas 20% were not. In relation to that, her experience with growth hormone substitution was very interesting. She had investigated a small group of truly growth hormone deficient children with Down's syndrome of less than 5 years of age. After the treatment they all were normocephalic. They had been growing more synapses, whereas a greater catch-up in IQ had taken place.

Dr. Wisniewski also mentioned a controlled study of 101 individuals with Down's syndrome and 80 non-Down's syndrome controls. In the last trimester of gestation she had already found subnormal brain weight, shorter frontal lobes, flatter visual centers, hypoplasia of the superior temporal gyrus in 30 - 40%, reduced cerebellum and brain stem, delayed myelinization maturation (visible on MRI as local dystrophy as well as maturation delay), basal ganglia calcification, hypopituitarism. Interestingly, 30 - 40% of the persons with hypoplasia of the superior temporal gyrus clearly appeared to be the ones with severe expressive language problems. She stressed the impossibility to achieve good expressive language in these individuals, independent of the type of intervention or therapy.

In another study as regards the brains of 100 individuals with Down's syndrome of over 30 years of age, the speaker had found many Alzheimer tangles, whereas only 20% of the persons involved had previously shown clinical symptoms of Alzheimer's disease. She presently finds very few adults with Down's syndrome getting clinical symptoms of Alzheimer's disease, if they are only active and working. In her opinion, the environmental factors are extremely important.

As regards the focus of future research the speaker mentioned genetic endocrine manipulation to enhance synaptic as well as neuronic survival.

A question in the discussion concerned the "upward development" of the group of Down's syndrome individuals in recent years. Dr. Wisniewski answered that:

1) previously, individuals with Down's syndrome were not exposed to education, etc. Only 20 - 30% were considered to be educable.

2) in the last decade, after a stimulating education, 70% of individuals with Down's syndrome are considered mentally retarded in the mild to moderate range, whereas 10 - 15% are only learning disabled. That is a tremendous difference due to proper education.

3) especially those who have been in institutions from their first day of life are mostly severely and profoundly retarded.

This very clearly shows the importance of the environmental factors.

**Cerebral pathology / Jesus Florez**

Prof. dr. Jesus Florez is professor at the Department of Pharmacology at the University of Santander, Spain. He made an attempt to bridge the gap between psychology and neurology. He started off by repeating the main points of the previous speaker.

All in all there is a lack of reactivity. Less information is received and generated, while, in addition, it is not conveniently processed. Yet, the formation of the brain is heavily dependent on the stimuli it receives. Whatever the child does in its first few years of life will shape its cerebral circuits. Because of that it is so extremely important what happens in these first few years. Yet, stimulation in later years remains useful. It is never too late. It should be the right type of stimulation, however.

The main problem area of children with Down's syndrome is the prefrontal cortex, used for decision making and the integration of information from elsewhere. Sequential organization of information is another task of the prefrontal cortex. Up to 40% of human brain weight consists of prefrontal cortex.

Prof. Florez listed some well-known aspects of children with Down's syndrome, which, according to him, are also caused by that prefrontal cortex problem: the inability to inhibit special conduct by themselves, the slowness in decision making, the reluctancy to change tasks, the lack of alertness, imitation and concentration, the difficulty to fix their gaze, the lack of initiative, the tendency to distraction and the absent-mindedness or hyperactivity with nothing in mind.

The sort of information that goes into a newborn brain is extremely important, the speaker emphasized. Stimuli even have to be intensified there until there comes a response. That means motivation for the parents. Without that the child with Down's syndrome is more passive. The extra stimulation is rewarding. The aforementioned behaviours are transient and early stimulation forces development. What is lacking is development of the attention control system. That again comes from the prefrontal lobes. However, that is the last area of the brain to undergo myelinization.

As such the child with Down's syndrome is much more limited by its environment than by its genes. The more possibilities it receives, the better its achievements are. Although, on the other hand, there is no reason to be too optimistic.

As far as future research is concerned, prof. Florez, considered the lack of knowledge on prefrontal lobes the main problem to deal with.

With regards to intervention programs, he recommended efforts to develop control mechanisms to be built into the programs.

**Specific medical pathology / Siegfried Pueschel**

Prof. dr. Siegfried Pueschel is Director of the Child Development Center and Professor of Pediatrics at Brown University School of Medicine, Providence, RI, USA.

Prof. Pueschel started off with the confession that he did not like to talk about all the abnormal features, with the danger of stereotyping Down's syndrome as a subspecies of the human race. He explained his practice of presenting a newborn child with Down's syndrome to its parents first and foremost as a human being. He then went on to list many of the features which are often seen in babies with Down's syndrome.

From there prof. Pueschel went on to explain common health problems in children with Down's syndrome. In the **neonatal period** both the gastro-intestinal and cardio-vascular problems might be life-threatening. In the third place there are the ophthalmological problems which the child has to be checked for.

The speaker emphasized that no form of treatment should be withheld that would be given unhesitatingly to other children.

As for the ophthalmological problems, prof. Pueschel stressed the importance of early diagnosis of congenital cataracts. If diagnosed too late the eye turns blind, while the operation is easy.

Congenital cardio-vascular problems have been the source of the high mortality of children with Down's syndrome until the mid-seventies. Prof. Pueschel strongly recommends every newborn child with Down's syndrome to be seen by a cardiologist on its first day of life. He mentioned having seen several children in heart failure during presentation in the weeks thereafter. An AV-canal is usually connected with a failure to thrive. Early pulmonary hypertension necessitates early surgery and in the meantime prescription of digitalis and diuretics. In most cases surgery brings about significant improvement.

During **the childhood period** there are the nutritional concerns. On the one hand does the child get the appropriate nutritional intake and on the other how to avoid overweight? For this purpose the Down's syndrome growth charts are of value.

Then there are the dental concerns: delayed eruption, abnormal sequence of eruption, partial adontia and microdontia, abnormally shaped teeth and crowding of teeth.

The permanent crowding is caused by the small oral cavity. Often it is necessary to expand the palate by means of braces, etc. Sometimes elements have to be extracted. Then there is the increased risk of gingivitis, seen as redness, an inflammatory reaction which can lead to loss of teeth. But all in all, there is no reason for not having a normal set of teeth in the end for a child with Down's syndrome.

There is an increasing frequency of youngsters with Down's syndrome with sleep apnea, which means a decrease of oxygen coming to the brain. It is caused by certain periods during sleep in which the child is not breathing at all. Change can be effected by correcting the sleep position (not on their back because of the tongue falling back and blocking the space of the air passage), removal of tonsils (if obstructing the airways) and decrease of overweight.

With respect to the thyroid function the speaker mentioned the increased frequency of auto-antibodies as well as hypo- and hyperthyroidism. Of these hypothyroidism is much more frequent. Two forms can be distinguished:

1) normal $T_4$ with very high TSH (compensated hypothyroidism) and 2) low $T_4$ with very high TSH (not-compensated hypothyroidism). In addition, prof. Pueschel said, there is a decrease of both $T_3$ and $T_4$ over time, which has to be checked periodically. An uncorrect funtion of the thyroid makes these persons even more retarded and ruins normal learning processes.

Further ophthalmological problems in the Down's syndrome population are: nystagmus (20 - 15%), strabismus (30%), cataracts (40 - 50% of older persons with Down's syndrome), keratoconus (5 - 7%), blepharitis and refractive errors.

Because normal visual acuity is extremely important for these children, particular attention should be given to thorough ophthalmological observation by a specialist with knowledge of children.

Common hearing problems are: conductive hearing loss (up to 80%), sensory hearing loss and mixed hearing loss.

The ear canal is often very narrow and obstructed by wax. The Eustachian tube runs more horizontally and drains less well. Fluid in the inner ear is a very good culture medium for bacterial growth, hence the high frequency of inner ear infections. Tympanometry is the recommended way to diagnose the presence of fluid. If there is fluid and inflammation antibiotics have to be applied. If necessary the inner ear has to be drained by tubes and if that doesn't bring about the necessary change, amplification has to be considered. The speaker stressed that hearing losses influence the psychological as well as the emotional development. In addition, a mild or moderate hearing loss leads to a reduced rate of language development.

The skeletal problems mentioned by prof. Pueschel were: atlanto-axial instability, loose ligaments in general, patella subluxation (in 5 - 7% of children) and hip dysplasia.

The speaker emphasized that symptomatic atlanto-axial instability can cause significant damage. He mentioned one child to have died of the effects of atlanto-axial instability. He repeated his well-known recommendation that all children with Down's syndrome should be radiographed at an age of 3.5 years.

In **adulthood** there are the related cardiac problems, such as mitral valve prolapse, present in over 50% of the population. From the point of view of nutrition there is the weight problem of many individuals with Down's syndrome. Finally, there is the possibility of Alzheimer's disease as well as other mental problems.

**Early intervention / Juan Perera**

Dr. Juan Perera is director of the Principe de Asturias Centre ASNIMO near Palma de Mallorca. The speaker outlined the environmental model of early intervention. He considered the process of development as a transactional process and the child as the active element. At first instance, he assumed the development of children with Down's syndrome to be quite normal but slower. Upon closing in, however, it was quite evident that the learning processes involved were quite different indeed. Down's syndrome itself influences the developmental process.

The social and emotional development are less involved and speech and motor development more. The problems in the motor development are governed by hypotonia, all the more so in the lower limbs, although the muscle tone improves over time.

Children with Down's syndrome are not as skillful as other children to use their environment. Down's syndrome babies and toddlers use their social skills to distract an adult from his goals.

Examples of atypical behaviour of children with Down's syndrome are: underuse of acquired behaviour, lack of motivation and impairment of eye contact.

As for the physical factors, the frequent hearing impairments were emphasized. In addition, there are other barriers that do not allow for information penetration. Transmission of information into the brain apparently is more difficult. As regards learning strategies the abnormal sensitivity of children with Down's syndrome to errors and mistakes were mentioned. Furthermore, even if the children know what to say they are often not able to express themselves. They frequently have immature, telegraphic speech. Perera stated that they did not talk in silence to maintain information. This means that the children do not exercise. However, their visual information processing is less decreased. Therefore, their teaching should underline the visual aspects. Because of that a compu-

ter is a very useful tool for them.

Many early intervention programmes were not as efficient as they should be. The solution could be found in syndrome-specific programmes, not one programme for every type of retardation.

Families experience less stress and are functioning more normally due to Down's syndrome self help groups.

Nowadays children with Down's syndrome are far more frequently only midly handicapped than severe as it was fifteen or so years ago.

He concluded that in Down's syndrome no intervention means regression. If youngsters or young adults with Down's syndrome are not supported throughout their entire life then all the work put in is wasted.

**Language systemic specificity / Jean Rondal**

Prof. dr. Jean Rondal is Director of the Laboratory for the Psychology of Language of the University of Liège, Belgium. He stated that there are no so-called pathognomonic symptoms in Down's syndrome. This means that there are no single symptoms that set Down's syndrome completely apart from other conditions. For example, articulation problems, very prominent in Down's syndrome, are not specific for the condition. From the point of view of etiology, trisomy caused by non-disjunctions is not specific for Down's syndrome, as there are also trisomy 13 or 18, etc. In addition, gene dosage effects occur elsewhere too. The speaker concluded that Down's syndrome is only specific at a more systemic level.

The speaker repeated that many Down's syndrome subjects have difficulties with phonics, while stuttering is also more prevalent (up to 30%). "Purely mechanical" sensory and motor problems help explain at least part of the problems: the buccal cavity is too small, the larynx too high in the neck, there is the hypotonia and often there are inflammations. As regards the voice quality there are different views. With the correct control group for comparison the difference vanishes, according to Rondal. Furthermore, there are auditory defects. Very frequently there are hearing losses, mostly of the conductive middle ear problem type, giving rise to losses of 30-50 dB bilaterally over the major speech frequencies.

Now, hypotonia is a problem for articulation. But, in addition to that, there is the motor planning and timing component. As regards timing of movements in general, one has to distinguish between the time to initiate a movement (the decision time) and the time taken to execute that movement (the movement time). In addition, the decision time itself is divided into the time that no electrical activity can be measured in the concerning muscle (pre-motor reaction time) and the time that electrical activity takes place, although the movement as such has not yet been initiated (motor reaction time). The delay in time to execute a movement, e.g. a movement of the muscles needed for speech, in particular concerns the pre-motor as well as the motor component of the decision time and not the movement time.

In many Down's syndrome subjects the grammatic function is distorted. Superficially, the development appears to proceed normally. A very global reference base for that is the mean lenght of utterance (MLU). There is a high positive correlation (0.88) between chronological age and MLU, like in the non-retarded population. However, when more sophisticated aspects are looked upon, the picture becomes different. For a certain part the morphosyntactic development of children with Down's syndrome is correct, e.g. as regards use of morphemes. However, in more complex sentences clear differences appear.

Mental age is a poor predictor of adult grammatical development. Lexicon development, on the other hand, is more in line with general cognitive development. The point of view according to which speech development in subjects with Down's syndrome is only delayed is not totally false, but, on the other hand, major parts of it do not conform to normal patterns of development.

Children with Down's syndrome are exposed to a language environment that does not differ, neither qualitatively nor quantitatively, from that of other children. As such the problems of children are not a product of the behaviour of their parents.

The belief today is that there are several genuine critical periods for language development. Articulation may be subject to a shorter time period than other aspects of language. About 20 years ago Lenneberg wrote that over 12 years of age no significant improvements might be expected anymore. This is not what the speaker found. According to him, there are still interesting developments in language possible after puberty at, say, 20-25 years, provided the relevant grown-ups are appropriately stimulated. So, language development need not be a matter of early intervention, but of continuous intervention instead.

The most serious language problems are usally associated with Down's syndrome. However, a part of these may not be purely due to the Down's syndrome condition itself. At present there is a beginning literature on cases of language exceptionality. Rondal discussed his own extensive study of a female with Down's syndrome, named Françoise, from chronological age 33-37. Her mental age is 5 years, 9 months, her verbal age 9 years, 10 months, while her IQ is 60 (WAIS). Her articulation is perfect. Her intonation is okay. She makes perfectly constructed, very long sentences (up to 58 words at a time), with correct syntax and correct morphemes. Her receptive language is even more expressive. All in all her language is virtually normal in its computational aspects. Its contents, however, are in relation to her cognitive development. Yet she has Down's syndrome. This example, together with the other examples, lead one to believe that the aforementioned phonological difficulties are not inherent to Down's syndrome as such, whereas the conceptual difficulties are. This brings back the argument put forward by Chomsky about the autonomy of grammar. In addition, there is much to say for a large degree of autonomy of phonological development. (This is the former so-called motor theory of speech). According to this, humans dispose of a special module concerned with motor invariance. Chomsky postulated that every human being was endowed with a kind of universal grammar, corresponding to a series of abstract parameters. And, in defense of that, how could Françoise have learned the kind of language she is using. Her immediate memory is not larger than 4 words! Her sentence span from memory is 14 words with occasional exceptions of 19-20 words. If there is genetically coded language information in all human beings then the same prerequisites hold for all individuals with mental retardation. All genes in Down's syndrome are normal! This means that whatever is contained in the human genome also is present in Down's syndrome. Therefore the basic problem in Down's syndrome is the **non-realization** of the potential. From there it was just one step to the so-called modular cognition approach. This means that distinct cognitive subsystems mature according to their own time tables. This model suggests that premature termination of the development should affect primarily those neural components and systems which would have matured after the premature arrest. It can be speculated that genetically coded grammar information will not be in a position to be realized in most subjects with Down's syndrome.

Rondal made a plea for a research orientation towards collaborative efforts of all concerned specialists, geneticists, phychologists, linguists, etc., into which effects are slowing down the development. The speaker prophesied that some time something significant about this could be done. In the meantime he considered enrichment of the child's conceptual environment well oriented and reasonable. However, systematic training in itself will only have limited effects on the computational components, because the natural development is tied to a priori information. Intervention has to contain early as well as continuous logopedic assistance. Furthermore, associative learning should continue to take place before one can install "a more frontal lobe oriented therapeutic approach".

**The associated Down movement in the USA / Donna Rosenthal**

Mrs. Donna Rosenthal is Executive Director of the National Down Syndrome Society (NDSS) in New York, NY, USA. She began with the statement that the trends in the associated Down's syndrome movement in the USA are much the same as in Europe. In the USA there are presently over 250.000 individuals with Down's syndrome with an average life span of 55 years. One in seven newborns with the syndrome has a life expectancy of over 55 years, she explained. Most of these individuals presently live with their families. Inclusion is an underlying trend in every aspect of their lives.

According to its director, the associated Down's syndrome movement in the USA is a typical "grass roots movement" with over 1.000 separate parent groups ranging from very sophisticated organizations with a paid, professional staff to very small groups entirely on a volunteer basis. All of these provide support and information. As the groups grow they all produce newsletters. Some sponsor programmes, medical clinics, social programmes and reach out to new parents, usually right in the hospital.

The parent movement is rapidly growing. Not only have the groups developed early intervention programmes, but they also give advice on that theme as well as offering social opportunities for teens and young adults. Because of this, the concerning families do not function in a vacuum. The role of the professionals is focused on research and service. Sometimes the professionals have to be convinced by the parents, while at other times it is the other way around.

The NDSS, as the national organization, was founded 13 years ago to educate the public and to support research. In its Committee professionals and parents are working together. All the information booklets produced by the NDSS are disseminated free of charge. At the present time there are 50.000 people on the NDSS-mailing list. Furthermore, it operates a full-time staffed toll-free hotline.

In the nine years that mrs. Rosenthal has been with the NDSS, the general population has become much more knowledgeable about Down's syndrome. A very important aspect of that has been the TV-series "life goes on" which now runs for the fourth year in a row on Sunday evenings. Initially, it hadn't that much of a success. Only after the NDSS has organized a press campaign it gained the popularity it now has, mrs. Rosenthal said.

Another important subject of the NDSS is the project "Child". It is a means to realize respite care for children with Down's syndrome aged 5-12 years once every six weeks with host families in the community. Due to this set-up everyone benefits, the child, the host family and the American tax payer.

In the U.S.A. early intervention is mandated by law. To achieve this, and other things, the NDSS testifid before Congress. For that purpose deputations of adults with Down's syndrome have travelled to Washington. They themselves are their best advocates.

A hot topic at the present time is the problem of adequate health care for many families with a child with Down's syndrome. Many insurance companies will reject them.

Mrs. Rosenthal explained that the NDSS itself does not receive any funding from the US-government, only for the research work it sponsors. So far the NDSS has had 15 scholars.

The work of NDSS is funded not by dues and fees. but mainly by voluntary grants from $5 upward. Therefore, the NDSS income consists of many tiny bits. Furthermore, there are bigger grants from other sources.

**Learning to read and write / Sue Buckley**

Sue Buckley is Director of the Sarah Duffen Centre and principal Lecturer in psychology at the University of Porstmouth, England. She began with the statement that children's minds are not determined by the structure of their brains. That only limits them. Very much on the contrary so, children's minds develop as they strive to make sense of the world around them, as they learn to understand, reason , remember and think. With this process children with Down's syndrome need more adult help. No two human brains are alike. Development is a dynamic interaction process. In what way are the processes of development altered in Down's syndrome, dr. Buckley wondered. She concluded that babies with Down's syndrome are distorting their own learning environment.

Once the children have a delay in language production their parents, as well as other adults in their direct environment, might no longer talk appropriately. The speech of the children might distort adult behaviour.

As for cognitive processes, the speaker considered speech to be the most powerful tool for cognitive functioning.

Dr. Buckley showed various pieces of video showing little toddlers with Down's syndrome happily reading flashcards. She emphasized that some of the children concerned find learning to read easier at three then at, say, eight or ten. But, she added, some late starters can also develop into very good readers.

The speaker pointed again to the importance of the discovery of the semantic errors of early readers with Down's syndrome. This means that the children were clearly responding to the meaning of the words on their flashcards. They were decoding these for meaning. So there was a direct acces from print to meaning. Only recently it became known that skilled adult readers do that all the time. Any motors skill one can think of will improve with practice. Reading means practice: therefore it will lead to improvement of articulation. She explained that only in the very exceptional cases that children with Down's syndrome cant't distinguish print, she and her co-workers use symbols.

In regard to writing the speaker recommended to practice writing alongside reading right from the start. This helps motor skills going and draws the child's attentions to the fact that letters make up a word.

**Variability / Cliff Cunningham**

Cliff Cunningham is emeritus professor of applied psychology in the Mental Deficiency Department School of Medicine, Nottingham University, England. He wondered how unique people with Down's syndrome really are. How does one change his way of interaction if one knows a person has Down's syndrome? He summarized the presentations on the congress with the key words variability and differences.

The history of the first 100 years of Down's syndrome was concerned with finding out in how far individuals with Down's syndrome were the same. Then came the research studies which compared Down's syndrome to non-Down's syndrome. Later again research was centered around children with Down's syndrome compared with children without retardation. This brought the speaker to the question whether Down's syndrome should be used at all as a collective term.

He went on by first describing in detail the many sources of variability in the development of children with Down's syndrome. But before doing so he referred to the strengh of their visual system, which seems to be quite unique. This is not found in other groups. It is one of few constants in Down's syndrome, in all the literature of the last 60 years. Therefore the teaching of reading, as proposed by the previous speaker, dr. Buckley, made an awful lot of sense, Cunningham said.

As the main sources of variability in his Manchester cohort he mentioned: families, genetic inheritance, environmental factors, secondary organic symptoms.

With regard to the environmental factors, he emphasized the transactional model of development. It is based upon the assumption: as a result of the interaction change accurs. The aim (of all education!) is to produce change. This transactional model is determined by: ecological system, culture, what is needed, environment, type of parent, context factors.

With regard to the family factors he made the differentations into: social-economic status of the parent, employment, presence of brothers and sisters etc.

As regards the facial features, according to the speaker, children with Down's syndrome are less likely to be involved in social situations, whereas they are discriminated by school psychologists in school placements, etc. It does have an effect. Appearance is furthermore correlated to stress measures in mothers and fathers, he said.

As "output factors" by means of which the variability could be measured Cunningham mentioned: behaviour, self-sufficiency, friendship scale, academic styles.

As result of his studies dr. Cunningham found the following factors to be associated with mental age: parental education and qualification (30 - 40% of variable!); gender: girls higher scores than boys; social class, 1, 2 and 3 higher than 4 and 5.

The speaker considered IQ to be a very good correlate of later development. IQ also is an indicator of inherited factors. Children with Down's syndrome from empoverished backgrounds are in double jeopardy. Because of this added deprivation problem, the biggest effect of early intervention can be achieved with these children in particular.

The best parents are controlled by their children. However, children with Down's syndrome do not get adequate control over their environment. Their parents are less well engaged. He mentioned six studies in which the mothers were trained in interaction styles because of which their babies did better.

Most studies find a gender effect: girls are somewhat more advanced than boys until an age of 15-16 years when the difference vanishes.

Children with Down's syndrome are often characterized by their lack of drive, their being slower to respond and more gentle and their being only contingent within a second or so. The latter pattern is well established at 6-7 months of age. The baby does something and the response of its parents does not show back to him understanding by his parents. This means that there is no semantic contingency from that time onwards.

Self-sufficiency appears to be highly related with mental age (over 50% of variability), but not to gender, parental education and social-economic status.

Academic attainments are strong predictors of mental age. For fathers

that felt to be in control of life, he had found their children with Down's syndrome have higher reading and writing scores.

As regards the type of school, mainstreamed children clearly showed higher academic scores, even when controlled for IQ or mental age and family background. Why was this so, Cunningham wondered? According to him, it was the pressure to drive the children into reading and writing in the mainstream and the absence of the tendency to wait for prerequisites, so prevalent in special school. Because of that, children in the latter type of schools were often simply not given the opportunity. In the mainstream also the other children were doing it, while the children with Down's syndrome were watching them. Typically, they have to be led and encouraged. The environment and the context will often do that. As such the type of school certainly had an influence.

Nobody has ever shown any feed forward effects. This means that if one stops the intervention no progress will be made anymore. Behaviours which are not maintained are lost. Generally speaking, the longer the child with Down's syndrome learns, the more he learns. With children with Down's syndrome in particular, one cannot simply trust a previously learned behaviour is still there because consolidation takes a long time. Lack of consolidation means at one time a certain skill is learned, somewhat later it is gone, then it is learned again, gone, learned, gone, etc. Therefore, too much hurry and too little overlearning leads to an instability of response and not to an automated skill behaviour.

Finally, the speaker gave a research focus for the next ten years: how much of the low arousal of children with Down's syndrome at the start of their lives originates in the first few weeks and is trained from them on?

*Erik de Graaf*

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.....

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.....
.....

Number of members: .....
Down's syndrome people's age (if personal affilation).....
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## GOALS AND OBJECTIVES OF EDSA

1. To spread throughout all European nations the principle that every person with Down's syndrome has the right to receive the health care and educational services demanded by his (her) condition, in order to achieve the best of his (her) possibilities.
2. To stimulate the implementation in each European country of a network of local groups, made up of parents and professionals. These groups should be able to better attend and resolve local needs, so that the families of every newborn with Down's syndrome may immediately receive the required support and advice.
3. To promote the principles of normalization in order to transform, humanize and dignify all human services upon which persons with Down's syndrome rely.
4. To encourage the development of programs and services that may be appropriate for persons with Down's syndrome.
5. To exchange information among the European countries on those programs that have proved to be effective. It is EDSA's conviction that the cultural pluralism of the European nations will enrich the personal and communal actions on behalf of the persons with Down's syndrome.
6. To introduce in all nations specific and comprehensive health programs for persons with Down's syndrome.
7. To encourage the constitution and convening of scientific groups, to share their study and research on:
   a) The biology of Down's syndrome and its pathological consequences.
   b) The mental development at different ages.
   c) Programs of education and intervention that are suitable for the specific conditions of each person with Down's syndrome.
   d) Integration in his (her) environment, in the community and at work.
8. To study and recommend legislation adapted to each European nation, in order to guarantee and ensure the services for the person with Down's syndrome during his (her) adult life.

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