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BRAINCHILD

The Official Publication of HKCNDP

香港兒童腦科及體智發展學會
The Hong Kong Society of Child Neurology and Developmental Paediatrics

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SPECIAL ISSUE ON VISION

CONTENTS

On the Cover is a very lovely drawing from
a child with visual impairment

The Hong Kong Society of Child Neurology & Developmental Paediatrics

BRAINCHILD – MARCH 2004 ISSUE

Message from the President

The current issue of Brainchild is devoted to Vision. Superior utilization of visual abilities in broad as well as focal attention is one of the biological characteristics which enables *homo sapiens* to claim its superiority amongst other members of the animal kingdom. Visual dysfunction is detrimental to the normal development of the child. Yet to clinicians, examination of the eyes and assessment of vision in a young child are always nightmares because of the demand for a skillful approach from the examiner and the need for good cooperation from the child. This Issue hopes to solve some of these problems through the sharing of experiences from experts in this area. We have Dr. Agnes Tse discussing "*Two Common Eye Diseases*" and Dr. Vishwanath et al presenting "*Achiasmia – A Review*". Both provide readers with clinical data on these medical problems. The paper on "*Visual Assessment of Pre-school Children*" by Ms Dilys Liu et al stresses on means for early detection while Dr. Iris Lau's article on "*Visual Impairment in Children – Developmental and Education Implication*" clearly delineates the importance of Vision on child development and education which are vital components in child health. Papers on "*Enhancing Vision Efficiency in Low Vision Pre-school Children*" by Ms Phoebe Yeung from Ebenezer School and "*Occupation Therapy for Children with Severe Visual Impairment*" by Ms Mandy Chui shed light on what we can do and what we can achieve for children with visual impairment. The contribution by Ms Clara Cheng et al on "*A Quest to Visual Function of Children with Cortical Visual Impairment and Severe Multiple Disabilities*" increases our understanding on magnitude of the problems. Dr. Iris Lau Kin Chun, Guest Editor for this Issue, is to be congratulated for compiling such a group of excellent papers covering areas of screening, assessment and rehabilitation of children with visual problems. I am sure readers will find the Issue both informative and interesting in reading.

At the Society level, we are pleased to report that the *Working Party on Epilepsy Surgery* has revolutionalized its protocol preparation, with clinical meetings of multidisciplinary and interdisciplinary expert teams discussing cases of refractory epilepsy which are potentially amendable to surgical intervention. It is envisaged that final working protocols will become clear as a result of these meetings of minds. The *Working Party on Cerebral Palsy* has just completed Stage I Survey to explore local epidemiological data for children with Cerebral Palsy. We are now analyzing the data and will soon be ready to launch Stage II of the Project targeting a wider coverage. The *Working Party on Specific Learning Disabilities (SLD)* continues to work on promoting public awareness, professional development, policy formulation and advocacy issues for individuals with SLD. We have successfully hosted two certificate courses with the Federation of Medical Societies of Hong Kong last year (one for Basic and one for Advanced Learners) which were well received by professionals with overwhelming attendance and favourable comments from participants. The Course on Children with Special Educational Needs (SEN) organized by our Society and sponsored by the Education and Manpower Bureau (EMB) of the HKSAR Government targeting over 2,000 school principals and teachers from primary and secondary schools was well attended despite a short interruption by the SARS epidemic. We plan to work further in cooperation with EMB, tertiary education institutions, teacher organizations in dissemination of knowledge and promotion of accommodation for children with SLD at school and in open high stake examinations. At the same time, the Working Party is still working diligently on *The Position Paper on A Service Model for Students with SLD in Hong Kong*, at the request of the Department of Health and the Education and Manpower Bureau of the SAR Government. We hope to have the Paper ready very soon.

This year is the Tenth Anniversary of our Society. The Council is planning a series of celebration programmes to commemorate this important milestone including publication of an *Anniversary Monograph*, hosting an *Anniversary Scientific Conference* in conjunction with the Society's 2004 Annual Scientific Meeting on Rehabilitation, organization of the *Anniversary Banquet*, and preparation of *souvenirs for members* (ties and scarves). An organizing committee has been appointed by the Council and all members of the Society are welcome to participate. We intend to invite guests and professionals both locally and internationally to share with us our local experiences in the subspecialties at this festive occasion.

On the promotion of our subspecialties of Child Neurology (CN) and Developmental Paediatrics (DP), the Society has been very active in supporting the Hong Kong College of Paediatricians which is the official academic body established by statute as a constituent college under the Hong Kong Academy of Medicine (*the Medical Registration Ordinance 1995*) responsible for training, education, quality assurance and CME of specialists/subspecialists. We have submitted our recommendations on training of both our subspecialties (CN and DP) in 1993 for publication of *the Guidelines on Postgraduate Training and Accreditation for Paediatricians* (1995), returned the completed *Questionnaire for Higher Training in Paediatric Subspecialties* (CN and DP) in March 2003, and now is ready to send in our *Application for Subspecialty Accreditation for Higher Training in CN and DP* by May 2004. Through these accreditation procedures, we sincerely hope to achieve official recognition of our subspecialties, to accredit qualified subspecialists in CN and DP, and to provide trainers for higher training in CN and DP as well as to reinforce the training programme for paediatrics (general paediatrics) which is always the foundation for all paediatric subspecialties. Most important of all, we hope to have accredited *Continuing Medical Education (CME)* and *Continuous Professional Development (CPD)* for our subspecialists and to have official channel of communication with corresponding subspecialists internationally for academic exchange, clinical collation and fraternity activities. Members please remain tuned for progress in this aspect.

Outside Hong Kong, we had a very successful symposium with *the Department of Paediatrics and Neonatology, Centro Hospitalar Conde S Januario, Macau* in 11-12th October 2003 on Child Neurology and Developmental Paediatrics in Macau. The Symposium was well attended by colleagues from both Hong Kong and Macau with active participation of experts (subspecialists) from the Mainland of China (Professor Wu Xiru from Beijing, Professor Jing Xingming from Shanghai and Professor Jin Jing from Guangzhou). We had presentations and discussions yielding good results and proposals for future activities. We are extremely pleased with the success of this Meeting and are encouraged to witness another Meeting being planned in Shanghai in May 2004 for further academic exchange, professional collaborations, and future cooperation in the subspecialty of Developmental Paediatrics. Internationally, our Society maintains good working relationship with the Asia-Oceanian Child Neurology Association (AOCNA) and the International Paediatric Association (ICNA). We are going to send out a delegation to the 8th AOCNA Congress in Delhi, India in October 2004 to show our support and to effect our academic participation.

In summary, our Society has been very active over the past few months as evidenced by the large multiplicity of aforementioned achievements. All these are fruits of good support of our members. For all your contributions, I say thankyou and I look forward to your further participation in the future.

I wish you all reading pleasure and best of health!

Dr. Chok-wan CHAN
Editor-in-Chief, Brainchild
President, HK Society of Child Neurology & Developmental Paediatrics

Two Common Eye Diseases in Childhood

Agnes Tse
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In this article, I will discuss two childhood eye diseases that can lead to visual impairment. They are Retinopathy of Prematurity (ROP) and Squint.

Retinopathy of Prematurity (ROP)

Retinopathy of Prematurity is a disease that occurs in very premature infants, usually with a birth weight below 1.5 Kg.

Embryonic development of retinal vessel

Retinal vessels start to develop at 16 weeks of gestation. They grow from the optic discs towards the periphery. At 9 months of gestation, the retinal vessels reach the far periphery of the retina and vascularization completes. If a child is born before complete retinal vascularization, the immature retinal vessels may develop abnormally and lead to ROP.

Mild ROP will regress spontaneously. Severe ROP will progress and lead to retinal haemorrhage and retinal detachment. The role of the ophthalmologist is to:-

1. Detect Threshold ROP (i.e. the threshold when treatment is indicated)
2. Give timely treatment (within 72 hours of reaching Threshold ROP) to
3. Prevent the ROP from progressing to retinal detachment

International classification of ROP

ROP is classified according to 3 features:

1. **Stage**
   ROP is divided into 5 stages: -
   Stage 1 – Demarcation line (Figure 1); Stage 2 – Ridge (Figure 2); Stage 3 – Extra-retinal proliferation (Figure 3); Stage 4 – Partial retinal detachment (Figure 4); Stage 5 – Total retinal detachment (Figure 5).

2. **Zone**
   The retina is divided into 3 zones, Zone 1 being most central and zone 3 being most peripheral. Retinal vessels grow from the optic disc to the retinal periphery. If ROP affects the more central zone, it signifies that the retina is more immature and the ROP is more serious. Therefore, zone 1 disease is more serious than a zone 3 disease.

3. **Extent**
   The retina is compared to the face of a clock with 12 clock hours. The extent of involvement of the retina by ROP is quoted as the number of clock hours involved.

Screening for ROP

All infants with birth weight < 1.5 Kg should be screened. The first screening is at 6 weeks after birth.

The infant's pupils are first dilated with 0.5% cyclopentolate and 2.5% phenylephrine eyedrops. The infant is examined in the nursery with an indirect ophthalmoscope. The purpose of the screening is to detect Threshold ROP (Stage 3) and treat it within 72 hours.

Threshold ROP

It is defined as:-

Stage 3 disease

Involving **Zone 1** or **2.**

Covering an extent of **5 contiguous clock hours** or **8 cumulative clock hours** (Figure 6).

Follow up regime

No ROP:

Observe; follow up every 2 weeks till the infant is 40 weeks post-conception.

ROP below the threshold level:

Observe; follow up every 1-2 week later till ROP regresses.

Threshold ROP:

Treatment within 72 hours.

Treatment of ROP

When ROP reaches threshold level, treatment has to be instituted within 72 hours.

In the past, treatment is with cryotherapy. Nowadays, it has been replaced by laser therapy, as this is less traumatic to the infant. Laser is delivered through a laser indirect ophthalmoscope by the ophthalmologist. Treatment is usually done in the nursery with the paediatrician giving sedation to the infant.

The rationale of laser treatment is to ablate the avascular retina to induce regression of the abnormal vessels. It has to be emphasized that the treatment window is extremely short. Threshold ROP can progress very rapidly to retinal detachment within 1 week. At that time, cryotherapy or laser treatment will be too late.

Cryo ROP study

The treatment results for ROP are very favourable. According to the Multicenter Trial of Cryotherapy for ROP, cryotherapy will reduce severe visual loss by half. (Laser treatment gives comparable results).

The CRYO-ROP study showed that 6% of infants with birth weight below 1.5 Kg would reach threshold disease. If untreated, 45% of these infants will end up with a vision of <20/200 at 3.5 years old. With treatment, only 26% will end up with a vision of <20/200.

Failed treatment

For a small percentage of ROP patients, ROP will progress despite treatment. They will end up having the following problems:-

1. Retinal detachment
2. Glaucoma
3. Flat anterior chamber
4. Phthisis (shrinkage of the eyeball)

Though retinal detachment in ROP can be treated with further retinal surgery, the visual prognosis is extremely grave.

Long term problems of ROP

If a child survives the acute phase of ROP during infancy, they still need to be monitored by an ophthalmologist as they have increased risk of developing the following eye problems:-

1. Dragging of the optic disc and macula
2. Late retinal detachment
3. Severe short-sightedness
4. Amblyopia
5. Squint

Summary

1. All infants <1.5 Kg birth weight are at risk of developing severe ROP.
2. Treatment of threshold disease will decrease severe visual loss by half.
3. ROP can progress very rapidly and the treatment window is very short. Therefore, the timing of referral for screening and the timing of treatment are very important.
4. Eyes that survive the acute stage of ROP need to be monitored for long-term problems e.g. late detachment, severe high myopia and squint etc.

SQUINT

What is squint?

Squint is defined as a misalignment of the eyes so that the 2 eyes are not fixing at the same object.

When one eye is fixing at the object of interest, the other eye may be: -

1. Deviated inwards (convergent squint)
2. Deviated outwards (divergent squint)
3. Deviated upwards or downwards (vertical squint)

What are the problems of having squint?

1. Squint may be caused by an underlying eye disease e.g. congenital cataract
2. Squint may cause lazy eye (The child only uses his fixing eye. The deviating eye is suppressed to avoid diplopia. This will result in poor vision in the deviating eye.)
3. Squint may lead to loss of binocular function
4. Squint gives poor cosmesis and has a negative effect on the child's self-esteem

How do we diagnose squint?

By torch reflex and cover test

Steps in treating squint

1. Exclude any underlying eye pathology. This should include a complete eye examination, including a fundus examination.
2. Check for any lazy eye and treat accordingly.
   If the deviating eye is found to be lazy and has poor vision, the patient should be treated by occluding the good eye until the lazy eye catches up the visual acuity of the good eye.
3. Aligning the eyes.

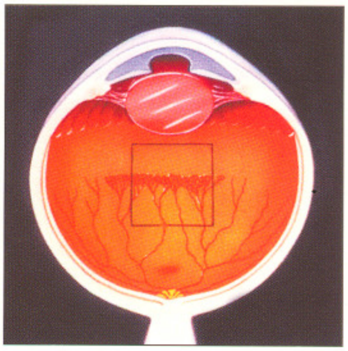

Figure 1: A faint white line is seen in the retina demarcating the vascularised retina from the avascular retina in the periphery.

Figure 2: The demarcation line develops into a ridge of tissue.

Figure 3: Fibro vascular tissue develops from the ridge into the vitreous.

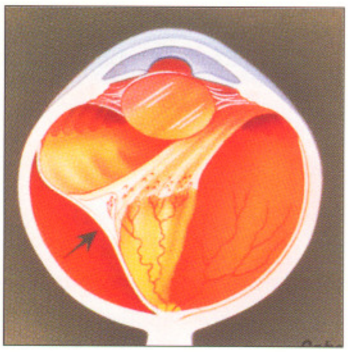

Figure 4: Part of the retina detaches.

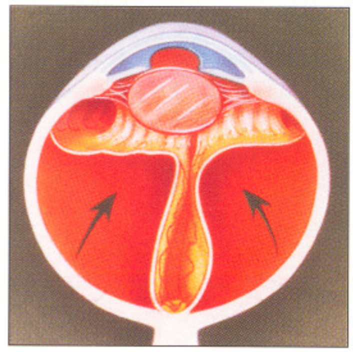

Figure 5: The whole retina is detached.

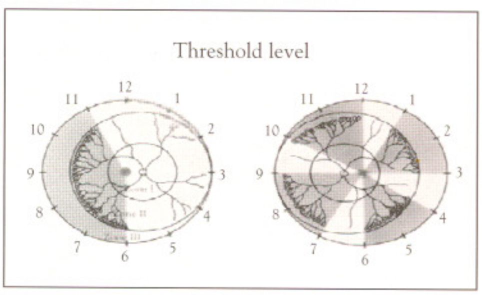

Figure 6: Threshold disease. Stage 3 disease involving 5 contiguous clock hours (left) or 8 cumulative clock hours (right).

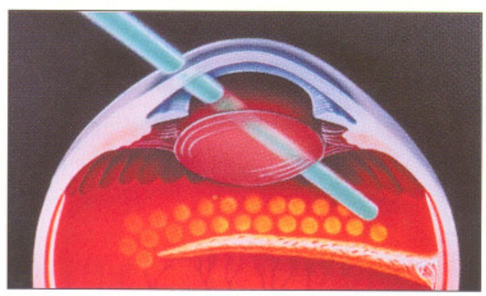

Figure 7: Laser treatment of the avascular retina for Threshold ROP.

Treatment to align the eyes

The method of aligning the eyes depends on the type of squint. In this article, only the most common types of squint will be discussed. They are namely:-

1. Convergent squint – Accommodative type (due to long-sightedness)
2. Convergent squint – Non-accommodative type (due to muscle problem)
3. Divergent squint – intermittent type

Convergent squint

Convergent squint can be caused either by long sightedness or muscle problem. These 2 types can be differentiated by checking for long sightedness after relaxing the child's eyes with pupil-dilating drops.

If a child has long sightedness, when he relaxes his eyes, objects are not in focus and look blurry. When he strains his eyes and accommodates, he will be able to focus and see more clearly. However, whenever he tries to accommodate in order to focus, the eyes will converge at the same time (accommodative convergence), leading to a convergent squint. In this child, if we give him a pair of long-sighted glasses, he will be able to focus clearly without straining and accommodating. In this way, the convergent squint will be eliminated. Therefore, the treatment for accommodative convergent squint is glasses. (Figures 8a & 8b)

If a child has convergent squint but does not have long-sightedness, the squint is due to a muscle problem. Treatment is by surgery, usually by recessing the medial rectus muscle of the eye. Surgery for convergent squint should be done before 2 years old and preferably before 1 year old. Otherwise, these patients will have poor binocular function.

Divergent squint

Most divergent squints are of the intermittent type. When the child exerts effort, he can control the alignment of his eyes. When he is tired or not concentrating, the squint manifests. If the divergent squint manifests frequently, surgery is indicated.

In a normal person, when the eyes are totally relaxed (e.g. when he closes his eyes or when his eyes are being covered), the eyes are not perfectly parallel. In most people, the eyes are slightly divergent (by a few degrees) in the relaxed state. In some people, the eyes are slightly convergent. However, once this person opens his eyes, he will realize that his eyes are not aligned and are looking at different objects. Instantaneously, the higher centers (fusion center) in his brain will direct his eyes to align and fuse the images in both eyes. Therefore, in everyday life, we will not see a normal person's eye deviating.

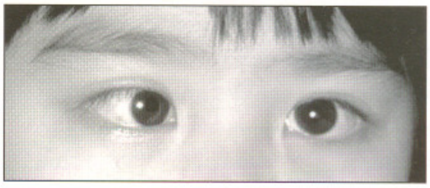

Figure 8a: Accommodative convergent squint before glasses.

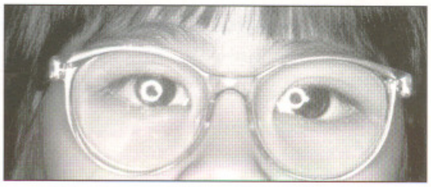

Figure 8b: Accommodative convergent squint after glasses.

Figure 9a: Divergent squint (before lateral rectus muscle recession).

Figure 9b: Divergent squint corrected after lateral rectus muscle recession).

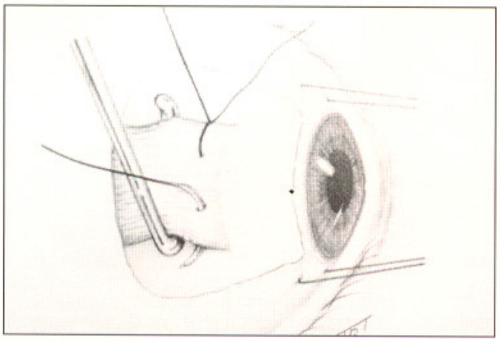

Figure 10a: Recession of the lateral rectus muscle. The lateral rectus muscle is first isolated.

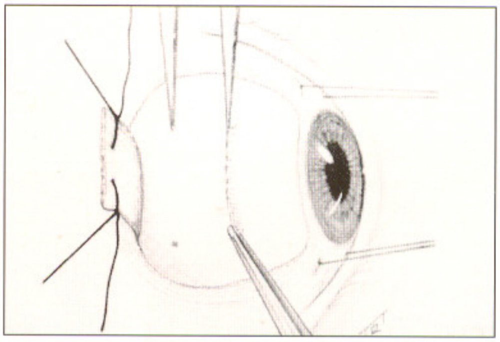

Figure 10b: The lateral rectus muscle is cut and recessed by a measured distance.

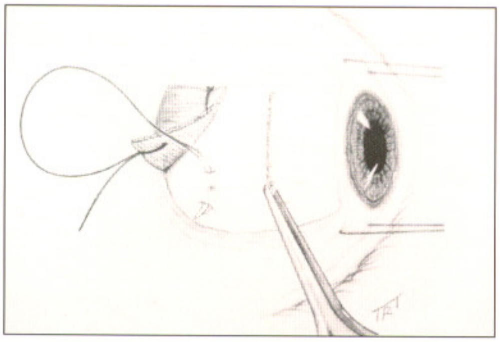

Figure 10c: The recessed muscle is sutured back to the sclera.

However, in a child with intermittent divergent squint, his eyes have a natural tendency to diverge out to a very large angle when his eyes are relaxed. When he opens his eyes, his higher center will try to pull the two eyes together. In view of the large angle, the higher center can only intermittently align the eyes. If the child is tired, we will see his eye diverging out. If the divergent squint only manifests occasionally, we can ask the child to do some fusion exercise to train his fusion power. However, when the squint manifests frequently, surgery is the treatment of choice.

The surgery for intermittent divergent squint is recession of the lateral rectus muscle. After recessing the lateral rectus muscle, the eyes will be in a near straight position even in the relaxed state. When the eyes open, the higher center will be able to align the eyes without difficulty. (Figures 9a, 9b, 10a, 10b and 10c). For this group of patients, because their squints are intermittent and not constant, surgery can be delayed until they are 4 years old, but before primary school age. Undue delay of surgery will lead to deterioration of binocular function. The surgical results for this group of patient are extremely good.

Summary

Squint is a misalignment of the eyes so that the two eyes are not fixing at the same object. An underlying eye disease can cause squint. Squint can also lead to lazy eye. Early treatment is indicated as lazy eye will become irreversible after the age of eight. For convergent squint due to long sightedness, using long-sighted glasses can align the eyes. For convergent squint due to muscle problem, the eyes should be aligned by surgery. For intermittent divergent squint, mild cases could be treated by fusion exercises. For severe cases, surgery is indicated. Surgery at the correct timing enables the patient to achieve the best binocular function.

Achiasmia – a Review

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Visual Sciences Unit, Institute of Child Health, London and
Department of Ophthalmology, Great Ormond Street Hospital for Sick Children, London

Introduction

Achiasmia is a rare condition of congenital absence of optic chiasm. It can occur alone or in combination with other midline anomalies. Historically there have been anecdotal descriptions of absent decussation of the optic nerves. For example, Lösel (1642) described the autopsy finding of achiasmia in a man who was hanged and anatomist Vesalius' described absent chiasm in a man he knew personally who had not suffered any double vision.[1] The typical VEP findings of this condition and its link with see-saw nystagmus have only been recently described.[2]

Case Report

A three months old female baby was referred with a history of pendular nystagmus, noticed by her mother from four weeks after birth. The birth and developmental history were normal. Her sibling of five years age was normal. On examination she could fix and follow light. There was a low amplitude horizontal pendular nystagmus. Vertical vestibulo-ocular reflexes (VOR) were intact, but horizontal VOR and optokinetic nystagmus were reduced. Some head nodding was also observed. Refraction and fundoscopy were normal. An Electroretinogram (ERG) and Visually Evoked Potentials (VEP) to flash and patterned stimuli suggested moderate to poor levels of vision with both eyes open. Additionally, an ipsilateral "crossed asymmetry" of VEP responses were seen suggesting a paucity of crossing fibres innervating contralateral hemisphere (Figure 1 and Figure 2). An MRI scan showed optic chiasm to be extremely small, but the remainder of the visual pathways and the rest of the brain to be normal (Figure 3).

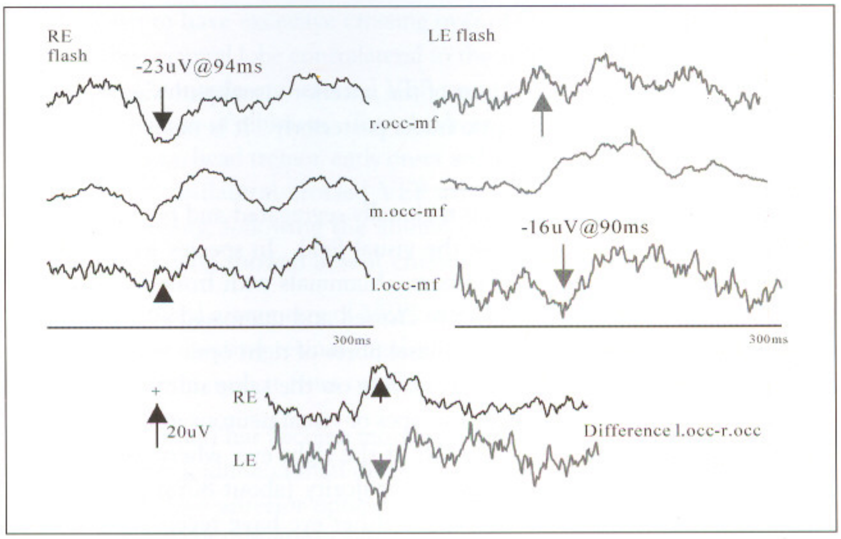

Figure 1: Monocular flash VEPs recorded from the patient at 2 years of age. Three occipital channels are shown for each eye. There is a prominent negativity on the ipsi-lateral channel to the eye stimulated i.e. for the right eye there is a prominent negativity over the right occiput and for the left eye there is negativity over the left occiput. When a difference of the two lateral electrodes is taken for each eye the trans-occipital polarity reversal or crossed asymmetry is emphasised.

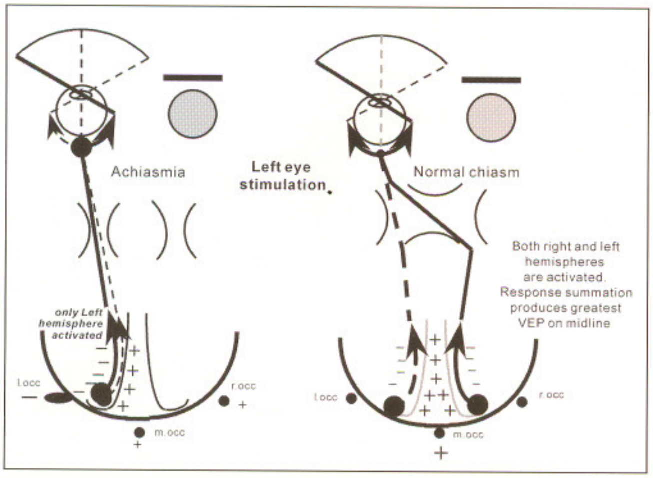

Figure 2: Schematic diagram illustrates the disproportionate innervation of the ipsilateral hemisphere in the absence of a chiasm. The cortical surface can be likened to a dipole sheet and the dipole projection will manifest as positivity over the right occipital electrode and negativity over the left, (ipsilateral) channel. Normally both hemispheres would be similarly activated and the summated response would be a positivity maximal over the mid-line.

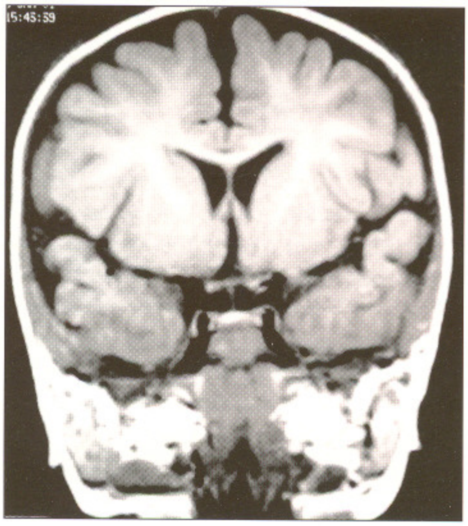

Figure 3: Extremely thin Optic Chiasm seen in the coronal MRI.

Discussion

The optic chiasm is the commissural part of the anterior visual pathway formed by converging of optic nerves anteriorly and diverging optic tracts posteriorly. It is named chiasm because of it's resemblance to the greek alphabet chi (χ).[3]

The visual information from the retina is regionally segregated and presented to the brain with individual fibres serving a designated part of the visual field. In species without binocular vision, the majority of the fibres decussate at the chiasm. Mammals with frontally placed eyes normally have an almost equal separation of the fibres into crossed and uncrossed at the chiasma. The fibres from each eye serving the binocular right field (nasal fibres of right optic nerve, crossing over to the left side and temporal fibres of left optic nerve, staying on the same side) bundle together to form the left optic tract and vice versa. There are two types of visual neurons in the primary visual cortex. Some are activated by signals solely from the left or the right eye, whereas others by signals from either eye. Animals, which are binocular, have a majority (about 80%) of neurons of the latter variety. Cells that can be driven by stimulation of either eye have receptive fields of nearly equal size and approximately corresponding positions in the visual field. The receptive field of a visual neuron is defined as that part of the visual field that can influence the firing of the cell. Visual association areas also have these binocular cells. The summation of the visual information from two eyes by these binocular cells in the primary visual cortex and visual association areas and sharing of information between the hemispheres via corpus callosum results in the single three dimensional visual percept of binocularity and stereopsis.[4,5]

Neuro-genetic studies with Drosophila, Zebrafish and mouse have helped to understand the development of optic chiasm and the pathogenesis of achiasmia. The growth cone navigation at the ventral midline is due to the differential ability of axons to respond to attractive and repulsive guidance cues provided by midline glia. Radial glia and early generated neuroepithelial cells located in the ventral diencephalon play an important role in this.[6] An important CNS ventral midline guidance cue is netrin-1, a diffusible protein secreted by ventral midline floor plate cells that can attract commissural axons. Netrin-1 and its receptor DCC ("Deleted in colorectal cancer") proteins are required for the normal development of retinal growth cone trajectories during chiasm formation.[7]

Neurodevelopment of this region is genetically controlled through transcription factors, regulatory proteins and signalling proteins. Genes encoding 14 transcription factors belonging to 6 different protein families are known to be required for development of mammalian eye. Pax genes are expressed in dynamic, spatial and temporal restricted patterns in the developing nervous system. Pax2 is a transcription factor gene expressed in the optic stalks reaching the brain and in a transverse band of neuroepithelium extending from one optic stalk to other.[8] Pax2 (paired box) and Noi (No isthmus) (zebrafish homologue pax2) are essential for axon growth within the optic nerve. Vax1 is a regulatory protein expressed by midline glia. It regulates the guidance properties of a set of anterior midline cells that orchestrate axon trajectories in the developing forebrain. Both Pax2 and Vax1 are essential for glial differentiation.[9] Shh (sonic hedge-hog) is a signalling protein expressed by neuroepithelial cells of ventral midline forebrain (future chiasmal region). Pax2 has an inhibitory role over Shh expression, evidenced by nonexpression of Shh in the region where the band (expressing Pax2) cuts the midline. If Shh expression is uninterrupted a proper chiasma is never formed.[8]

The discovery of the condition of Achiasmia in humans is an interesting story. Autopsy on Belgian sheepdogs with pronounced congenital nystagmus, had revealed complete failure of optic nerve decussation. Ocular motility studies in these animals, with this known absent decussation, had showed elliptical nystagmus, see-saw nystagmus and horizontal nystagmus.[10] Hence a link between see-saw nystagmus and achiasmia was established.

Albinos are known to have excessive crossing over of fibres at chiasma. This leads to excessive electrical activity at the occipital lobe contralateral to the side of the eye stimulated during monocular VEP tests. Evidence of "contralateral crossed VEP asymmetry" related to this excessive decussation is considered pathognomonic of albinism. Two children[2] with clinical features like reduced distance acuity, alternating esotropia, head tremor, early onset see-saw nystagmus and intact but poorly defined foveal reflexes, but with ipsilateral crossed VEP asymmetry, not consistent with albinism, were investigated by MRI imaging, following the animal evidence of association of see-saw nystagmus with achiasmia. MRI imaging showed absent chiasm but with no other neurological abnormality.

Thus "ipsilateral crossed asymmetry of VEP" was linked to achiasmia. In 1994 Apkarian et al[2] coined the term "non-decussating retinal-fugal fibre syndrome" to this newly discovered isolated congenital anomaly to distinguish it from other conditions of which achiasmia may be a part.

Since then, the condition has become more widely recognised. Chiasmal anomalies have been described in patients with midline cranial defects like septo-optic dysplasia, or midfacial clefts, or basal encephalocoele.[11] Also anterior optic pathway anomalies like hypoplasia or aplasia of optic nerve[12] and anophthalmia are sometimes associated with chiasmal defects.

See-saw nystagmus was initially thought to occur in association with isolated interruption of chiasmal fibres. However, it has been reported with chiasmal hypoplasia with midline anomalies.[11] Vertical and torsional eye movements are controlled by central otolithic and vestibular pathway at the diencephalic-mesencephalic junction. Hence, it is no surprise to find this with midbrain anomaly. See-saw nystagmus has also been reported with severe visual loss of cone rod dystrophy and some types of Miners nystagmus. Pendular nystagmus has been suspected to occur when central feedback circuits passing through chiasma to inferior olivary nucleus are disrupted causing synchronous oscillations of floccular purkinje cells resulting in nystagmus.[13]

What effect does this failed decussation have on visual quality? One study[14] showed, oculomotor instability resulting in nystagmus apart, fine vernier acuity, texture perception, three dimensional object perception using non-stereo depth clues were normal. Contrast sensitivity and velocity judgement were affected, but thought to be due to oculomotor instability rather than miswiring per se.[14] This behaviourally normal vision demonstrates the plasticity of retinal projections and cortical organisation. Absence of stereopsis highlights the need for retino-geniculo-cortical input from corresponding points rather than just midline interaction say via corpus callosum.[3] Acquired chiasmal defects, in contrast, develop bitemporal hemianopia because of disruption of nasal fibres. Therefore intact temporal fields would rule out an acquired defect.

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Visual Assessment of Pre-school Children

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Vision, together with other sensory and motor functions, plays an important role in the development of infant and pre-school children. Children develop by exploring and learning through their interacting with the environment. They see, they hear, they touch and they imitate. Through this process, they develop. Vision is a vital source of information about the environment: colours, form, shape, contrast, differences etc. This information is essential for the normal development of children. It is this information that enables them to acquire knowledge about the world around them and to get advances in their cognitive abilities. This is especially important for pre-school children because of the rapid cognitive development during this period of time. If vision is defective and is left undetected, it may become unrecoverable and may lead to delay in development when compared with their peers.

Assessing the visual functions of pre-school children is challenging, especially those who are identified or suspected to have other developmental problems. The presence of developmental problems may not interfere the normal development of visual function in general, but their presence may, in some cases, affect the result of the assessment. Having the knowledge of the normal development of visual functions helps to identify the visual problems that the child may have so that early detection provides a better prognosis in treating the problems.

Common Presentations of Visual Problems

Children with defective vision may present with different signs and symptoms depending on the causes and the severity of the defects.

The most common presentation, by far, is an abnormal head posture. Abnormal head posture may take the form of a face turn, head tilt, chin elevation or depression or their combinations. Children adopting an abnormal head posture may be for a clearer vision as in case of nystagmus, wider visual field in the primary position in case of bilateral partial ptosis, single vision in case of paralytic squint, which will, at times, elicit double vision.

Squeezing of eyes is sometimes found in children with reduced visual acuity. They squeeze their eyes to create a pin-hole effect for a clearer vision.

Wandering eye movements or even lack of eye contact may be seen in children whose vision is severely impaired.

The presence of white pupil in one or both eyes needs to be attended to with urgency because it may be a sign of cataract, retinal detachment or even retinoblastoma.

Visual Assessment

Visual assessment of pre-school children may include the following areas:

- Visual acuity
- Visual field
- Colour vision
- Binocular single vision

Visual acuity

Visual acuity refers to the spatial limit of visual discrimination. It is a highly complex function consisting of:

1. The ability to detect an object in the field of vision.
2. The ability to separate two critical elements of a test pattern.
3. The ability to recognize a symbol.

Visual acuity is the measure of the threshold of these abilities. Vision is one of the senses that children use to learn during their developmental age. The level of visual acuity reflects the intactness of the visual system. Decrease in visual acuity indicates that there is somewhere wrong in the visual system. If left unattended, information received through the visual system will be inadequate for the child to develop appropriately.

At birth, infants have all the eye structures necessary for them to see. The visual system is not fully developed yet; therefore the vision of an infant is relatively very blurred. As they grow, visual acuity develops dramatically. It is always difficult to truly represent the visual acuity of children under the age of 2 in Snellen equivalent due to the nature of the tests employed.

However, visual acuity may reach adult level by the age of 2 to 3.

Assessment of visual acuity:

a. Fixation behaviour:

This is not a measure of the visual acuity of the eye. It is rather a gross assessment of the vision by assessing the response of the child either binocularly or monocularly and the ability to fixate on a target of interest. 100's & 1000's, rolling ball and mounted ball are tests of this type.

b. Preferential looking technique:

It is based on the principle that when two targets, one is plain without any details and the other with details, are presented, the infant and young child will prefer to look at the target with details as long as the details can be resolved. Cardiff Acuity Test (Figure 1) is designed using this technique.

c. Matching tests:

When the child is old enough and has the ability to perform matching, Kay's picture, Ffook's Symbols (Figure 2), LEA Acuity (Figure 3), E-cube, Sheridan-Gardiner test (Figure 4), logMAR crowded test or, in some cases, even Snellen test type can be used. These tests can be done either by matching or by verbal response. For normal children beyond the age of 4, verbal response is usually possible.

Visual acuity is usually recorded in Snellen's notation of metres (e.g. 6/6) or feet (e.g. 20/20).

Visual fields

Visual field is that part of space in which objects are simultaneously visible to the steadily fixating eye. It can reflect the integrity of the visual pathway. Visual field develops in line with the development of visual pathway and expands gradually in size in all meridians to the extent comparable to that of an adult at the age of about 2 years old.

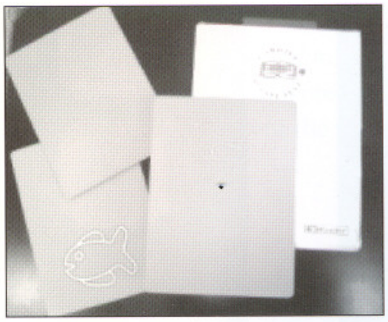

Figure 1: Cardiff Acuity Test

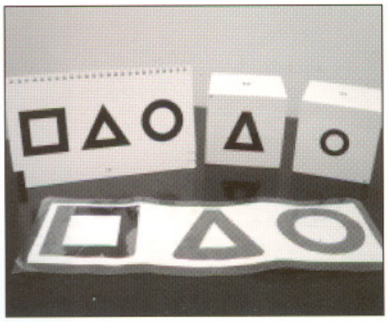

Figure 2: Ffook's Symbol Acuity Test

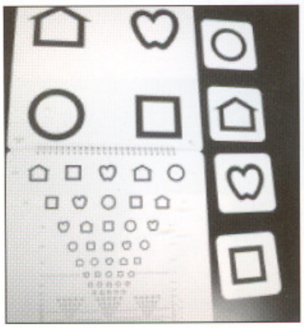

Figure 3: LEA Chart Acuity

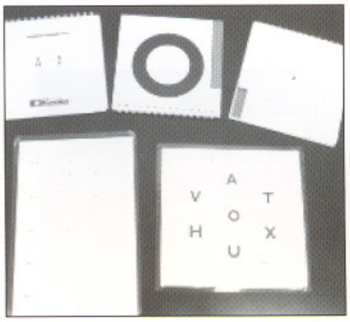

Figure 4: Sheridan Gardiner Acuity Test

Objective of the visual field assessment is to assess the integrity of the visual pathway. Clinically, it can be assessed statically and kinetically with very sophisticated machines, but to those pre-school children, these machines seem to be too frightening for them. It is therefore performed with them fixating an object of interest in front of them while the examiner slowly introduces a stimulus to the child's lateral side from behind. The movement of the child's head is observed. This is usually done binocularly in four quadrants as it encounters less resistance from the child.

Colour vision

Approximately 8% of children have an abnormality in colour perception. Most of them are boys. Among them 2% are chromatic defective and the remaining 6% are chromatic anomalous. Most of the colour vision defects are congenital. Acquired colour vision defects are usually associated with defective visual acuity or visual field loss.

Testing of colour vision in pre-school children has become essential in terms of learning and child-teacher relationship. If the defect can be identified early and teachers are aware of the situation, the unnecessary misunderstanding of not following instruction, especially in colouring tasks, can be avoided.

There may be a collection of colour vision tests designed to test the colour vision of young children, such as Make Easy Color Test (Figure 5), City University Test (Figure 6) and Ishihara's Test (Figure 7). Whatever tests one intends to use, it is important to select the one that is appropriate to the ability of the child for a reliable response.

For children younger than 3, it may not be possible to get a subjective response at all.

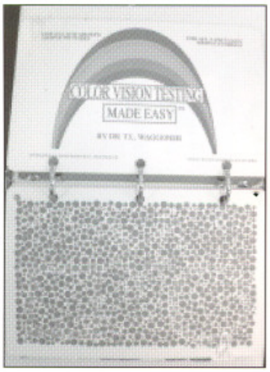

Figure 5: Make Easy Color Test

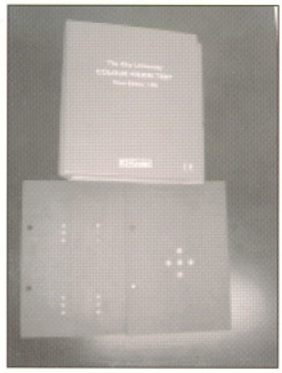

Figure 6: City University Test

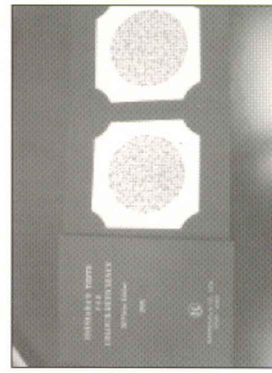

Figure 7: Ishihara's Test

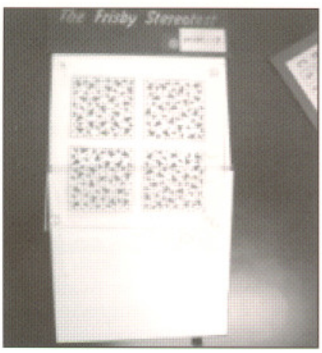

Figure 8: Frisby Stereotest

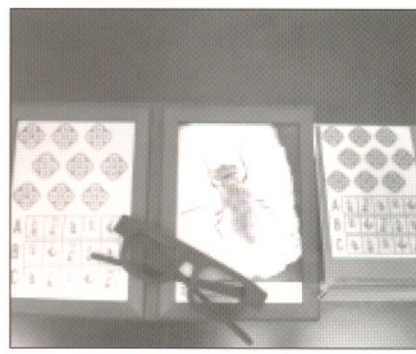

Figure 9: Titmus Fly Stereotest

Binocular Single Vision (BSV)

Binocular single vision is regarded as an ability to fuse as well as an ability to have stereopsis. For BSV to be developed normally, the sensory and motor systems of the eyes must develop in the way that they work hand in hand with each other, so that

1. The field of view is enlarged by transforming the field of vision into the field of fixation.
2. The object of attention is brought onto the foveae.
3. The two eyes are properly aligned at all time.

The two eyes work as a pair at an age as early as three months old. This implies that fusion is present at the early stage of life and is getting mature as the child grows. During this maturing period, any sensory and motor obstacles to fusion may disrupt the proper alignment of the two eyes and a manifest squint results. Sequelae of a squint include loss of depth perception, suppression and amblyopia of the squinting eye.

Adults may learn to use monocular clues for the estimation of relative distance of visual objects. Young children may encounter difficulties in the course of learning in school.

Stereopsis can be assessed by using various stereotests such as Lang, Randot, Random Dot, TNO, Frisby (Figure 8), and many others.

They measure stereoacuity from 2000 seconds of arc to 20 seconds of arc.

Clinically, cover test is used to detect the presence of a squint. The child is attracted to fixate on an object at near (1/3m), distance (6m) and far distance, when indicated, while the eye is covered and uncovered alternately. If a squint is present, whether it is constant or intermittent, further investigation by an ophthalmologist to rule out the presence of organic causes is required.

Conclusion

Children see and learn through the sensory and motor systems of their two eyes. Any interference to the development of these systems will create obstacles to fusion as well as the normal development of their vision. Impairment of vision will affect the development of a child in general. Identification of such obstacles at an early age can give a better treatment outcome both for the development of the vision and the development of the child.

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Visual Impairment in Children – Developmental and Educational Implications

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Vision is the most important sense to perceive the world around us. It is even more significant during the infancy period when the infant depends on vision substantially to learn about her environment Severe visual impairment therefore poses a developmental emergency in infancy, where early recognition and intervention is crucial for optimal development.

Definition and Epidemiology

Visual impairment (VI) is defined mainly by visual acuity and visual field defects. However, the presence of significant eye movement abnormalities also impairs the visual function and should be taken into consideration when assessing the visual difficulties. The World Health Organization[1] has defined varying degrees of visual impairment according to the best corrected visual acuity and the field of vision (see Table 1). It is worth noting that the Hong Kong Rehabilitation Service and Special Education Service follow a slightly different terminology. Nevertheless, the eligibility for service provision follows the same quantifiable disability.

Around two thirds of patients with visual impairment have multiple impairment including cerebral palsy, epilepsy, mental retardation or hearing impairment.

Table 1: Classification for visual impairment

Snellen Equivalent (best corrected visual acuity)	6/7.5	6/18	6/60	3/60	1/60	LP
Clinical			Count fingers <6m	Count fingers <3m	Count finger <1m/ Hand movement <5m	No light perception
WHO ICD-10 (1992)	(Slight impaired vision)	*Cat. 1* Moderate LV	*Cat. 2* Severe LV (or visual field ≤20° around central fixation)	*Cat. 3* Profound LV* (or visual field ≤10° around central fixation)	*Cat. 4* Near total Blindness	*Cat. 5* Total Blindness
Special Ed HK		Mild LV (Eligible for service e.g. SEN service)	Mod. LV	← Severe LV →	← Severe LV →	Totally blind
			← School for Blind →	← School for Blind →	← School for Blind →	← School for Blind →
Disability allowance (SWD, HK)				← Worse than 3/60 in the better eye →	← Worse than 3/60 in the better eye →	← Worse than 3/60 in the better eye →

* *LV = Low vision*
LP = Light perception

In Hong Kong, we do not yet have a vision impairment registry on the prevalence of visual impairment. The data from Rehabilitation Services may grossly underestimate the prevalence as cases are mainly reported through the Schools for the Blind. Moreover, formal visual acuity assessment cannot be conducted in children with cognitive impairment. In developed countries, prevalence estimation in children is between 0.8 to 1.8 per 1000 for visual acuity of 6/18 or less, and around 0.2 per 1000 for acuity of 6/60 or less.[2,3,4]

Pattern of Development

Observations have shown that the constraints on early development imposed by impaired vision are far-reaching and complex.[5,6,7] The effects depend on the integration and interpretation of input from other senses. Areas affected include the development of emotional bonding, personality and self-concept, social interactive skills, sound and tactile localization skills, fine motor and locomotor competence, object permanence, language and other cognitive concepts.

Compared to children with some functional near vision, children with profound visual impairment (those who show no visual awareness of object at close distance) have a higher incidence of poor developmental outcome. Up to 30% may have plateauing or loss of cognitive and language skills followed by an extremely slow rate of learning. Behaviourally, these are intensifying negativistic behaviour, failure of social communication, social relating, increasing self-stimulation and stereotypies.[8,9,10] These regression and autistic-like behaviours occur mostly during the second or third year of life. Other risk factors are the male sex and an increasing number of brain lesions.[11,12] This phenomenon is partly attributed to the disruption of referential communication, in the sharing of interests and attention to objects with an interacting adult.[12]

Without functional vision, children may not develop at the same rate or the same sequence as sighted children. They encounter great obstacles, especially in early life. Many skills and behaviours have to be learned in a different way. However, once these hurdles are overcome, the intellectual potentials can be achieved as for a sighted person.

Social hurdles

One of the most significant achievement in the first year of life is the attachment of the infant to parents, forming the basis for emotional security and development. In a visually impaired child, the lack of eye contact, the altered response to touch or voice like startle or stillness, greatly hamper the emotional feedback from the parents. Early guidance to parents on ways to interact with the child is crucial to healthy emotional bonding.

The child with VI also has difficulty reading other's emotion, gesture and body language. This is a major disadvantage to social interaction and communication. Skills such as turn taking, careful listening to others has to be specifically taught and practised.

Cognitive hurdles

Visual impairment affects how children learn, including *object permanence, parts to whole learning and categorization*. Object permanence is the concept that something continues to exist even when out of sight or touch. Children with VI need extra help to feel for dropped objects, to associate objects with consistent labels and use.

While sighted people see the whole object first, children with VI are limited to what they see or feel at any one time. They need to construct the image from pieces of information. They rely a lot on second hand experience or verbal prompts. Learning needs to be structured and well sequenced to create meaning and understanding for the child. This involves integrating the senses through touch, sound and previous experience on similar objects.

Children learn to categorize or sort objects by physical characterisitics, such as colour and shape. Then they sort by group, function and association, like things that you need to go to school. Through categorizing things, one can organize the thinking and build on existing concepts. Children with VI have difficulties understanding the similarities and differences of things in their environment. For example, in the process of learning about pets and animals, we need to verbally point out the similarities and differences as well as letting the child feel the different qualities.

Language hurdles

Like sensorimotor and cognitive skills, even some residual functional vision (like seeing objects at near distance) can make a big difference to gaining concepts and skills. A blind child can usually imitate vocal sounds and words. However, they may have problem in understanding the meaning, association and context in using the words. Their expressive speech is usually more advanced than verbal understanding. Words and phrases may be parroted out of context. Pronouns such as "I" and "you" can be mixed up.

Learning through using real objects and adding on previous experiences may help these profound visually impaired children to acquire word concepts. Enhanced exposure to surroundings and interaction with people may help to relate experiences and events.

Sensory hurdles

Vision plays a major role in integrating our senses as it takes all separate pieces of sensory information and build them into a whole. In the absence of vision, this process is much harder, as the senses of smell, touch, hearing and taste may not come in on a constant basis.[13] Sounds may differ according to the distance and the presence of other ground noises. Objects can be touched only if they are within reach. Intersensory integration is more difficult to achieve without vision, and the child should be guided through exploration with different sensory input. Care needs to be taken not to overload them with multiple stimuli but to present them in sequence that the child can perceive and understand.

A common problem encountered by children with little light perception is sleep disturbance. The confusion between day and night makes it difficult for the circadian rhythm to establish. Also, these children tend to be less active physically and may not be ready for sleep at bedtime. A consistent daily routine is essential to keep the child active in the day. Reinforcing a bedtime routine will also help in setting the biological clock.

Motor hurdles

Most of our motor skills are learned through watching and imitating others. Children with little useful vision have difficulty picking up skills such as crawling, walking, jumping or skipping.

Likewise, fine motor manipulation and exploratory skills are restricted and delayed. A systematic, hand-over-hand technique in manipulation and movements will help the child overcome these obstacles. Readers can refer to the paper by our occupational therapist for details.

Children with Cortical Visual Impairment

Cortical visual impairment refers to impaired vision from lesions of the optic radiations, striate cortex or occipital cortex. It has emerged as the leading cause of visual impairment in developed countries, reported in 40-50% of visually impaired children.[4] With the predominant causes being perinatal or postnatal hypoxia, head trauma and cerebral malformations, co-morbidities are common. These include cerebral palsy, epilepsy, mental retardation and hearing impairment.

However, one remarkable aspect about cortical VI is the retention of residual vision which often improves with time.[14,15] Yet careful observations must be made with regard to any defects in visual field and ocular motility. Moreover, children with cortical VI show variable visual efficiency and often have short visual attention. They tend to function best when relaxed and away from visual or sound distractions. Targets need to be presented against plain contrasted background.

Educational Implications

A severe visually impaired child has a wide range of educational needs including training in listening skills, tactile discrimination, orientation and mobility, literacy and social adaptation. Special curriculum and learning materials, coupled with specially trained teachers in the use of Braille, low vision aids, orientation and mobility are essential. Unless these resources can be provided sufficiently in the regular mainstream schools, the severe visually impaired child may benefit more from structured training programs in a special school. Integration into mainstream education may be considered only when the child has acquired basic skills and is able to adjust to the less intensive support in regular schools.

For children with multiple handicaps such as physical and mental impairment, the curriculum needs to be specifically tailored to the abilities and limitations of the individual. The areas of potential development include independency, communication, physical mobility, social skills and emotional development. Depending on the major handicapping condition, the child may benefit from training in special schools for physical, mental or visual handicap.

For children with mild to moderate low vision, integration into mainstream schooling is a major issue when the child approaches school age. Adequate accommodation and active intervention on the child's visual, learning and mobility skills are needed to ensure the psychosocial well-being of the child. Class teachers need regular support from specially trained vision teachers (from the outreach program of the Ebenezer School for the Visually Impaired) to provide advice, some direct teaching and modified learning materials.

Conclusion

The difficulties faced by the family and the child with severe visual impairment are wide ranged. Early detection and diagnosis can clear the uncertainties in the family with early adjustment and recognition of the challenges ahead. Urgent support to the family members and structured training is crucial to the healthy growth in the child both psychosocially and intellectually.

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Enhancing Vision Efficiency in Low Vision Pre-school Children

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Research findings have shown that with a planned learning programme for promoting visual functioning efficiency, children with low vision can develop their visual functioning ability to the highest potential (Barraga & Morris, 1980). Barraga and Morris also advocate that early intervention is important because the "early years are the most crucial years for sensory stimulation and perceptual development" (p.66). If the child lacks visual stimulation at the infant stage, the retina and the visual pathway to the brain may not develop and the visual receiving area in the brain may also be affected.

For infants, vision assessment and training can be in the form of visual stimulation. Assessment and training are often interconnected. They are "completely dependent on each other and cannot reasonably take place without the other" (Best, 1992, p.136). By giving visual stimulation, we can observe the visual response of the baby. At the same time, the baby is learning to see while being given such stimuli. Parents or caretakers can use torchlight, toys and daily objects of high contrast as stimuli. With newborn babies, we can start with black and white patterns because black and white contrast attracts them most. Later, we can use brightly coloured objects. A good make-up face of mummy is most desirable for a peek-a-boo game. Bringing the child's hands to midline for him/her to see and play with is also important. At this stage, visual awareness, visual attention, tracking and reaching out for objects seen will be the main focus.

"When the vision stimulation activities are completed, the child will be ready for visual efficiency activities" (Jose, 1983, p.401). For toddlers, rolling balls, treasure hunts, simple matching games using three dimensional objects and simple pictures can be used to further enhance visual functioning and develop visual perceptions. As toddlers are more locomotive, we should encourage them to use their vision for coordination of motor system with vision system. We can encourage them to use their vision to guide orientation and mobility, and to pick up small objects such as jellybeans to promote eye-hand coordination. The miniature-toys test and the graded ball test in the STYCAR Procedure (Sheridan, 1976) can give us a rough idea of the child's level of functional vision.

Between age three and four, there is rapid expanding of visual development while "organization of visual perceptions overlaps and merges with specific functional refinements" (Barraga, 1980). When children enter kindergarten, some more formal assessment kits such as the Look and Think Checklist (Tobin & Chapman, 1972) or the Diagnostic Procedure Assessment (Barraga, 1980) can be used to assess the child's visual abilities. After the assessment, the teacher can then write an individual training programme to focus on areas of weakness. More emphasis can be put on perceptual vision. Activities include discrimination and recognition of more details in pictures and photos, matching and naming shapes and colours, understanding pattern and symmetry etc. We should also prepare the children to read and write in the older age by giving them exercises like symbol tracking, symbol discrimination, visual memory games and most importantly, eye-hand coordination exercises like pencil maze and colouring. We can encourage children to explore small objects with simple magnifiers or a CCTV. Telescopic aids can also be given to promote interest in things and activities in the distance.

There are several things parents, caretakers and personnel working with low vision pre-school children should note:

It should be remembered that assessment and training of vision should always be in the form of games or pleasurable activities. Chen (1997) points out that it is more desirable to use natural and comfortable ways to help young children to learn to look. Lindstedt (n.d.) stresses that using vision should be taken as a positive experience and as fun. Sometimes parents or teachers may emphasize too much on training and it may create tension on both the adult and the child.

Owing to their restricted vision, low vision children will have to make great effort to see. Some children may need longer time to look and respond to visual stimuli. Sometimes, ability to perform some visual task may remain primitive for a long time before it is performed when least expected (Barraga, 1980). In order to encourage them, ample time should be allowed and recognitions and praises should be given for a small success or effort made. Meanwhile, we need also to perceive the child's limit of visual capacity. "If vision is very low, undue emphasis on vision may do more harm than good" (Lindstedt, n.d. p.1)

When designing a training programme, we should bear in mind the age, mental ability, motor development and motivation of the child. The efficiency of visual function would be limited by a person's cognitive and perceptual development and, on the other hand, promotes such development in an integrative way (Barraga, 1980). While providing vision training, we should also consider the holistic development of the child. Vision impressions "have to be combined with impression from other senses, linked up with motor functions and processed conceptually and emotionally" (Lindstedt, n.d. p.1)

Involvement of parents in assessment and training is crucial. Parents are the ones who know their children best and they can provide valuable information about the children's use of vision in natural situations (Chen, 1997). As children with low vision need ample visual stimulation, parents' follow-up and reinforcements of the skills learnt in the center or at school can help the child to apply the skills to their daily life.

As many areas of development can be affected by impaired vision, techniques for vision enhancement and vision substitution should be employed to support the early development. (Hyvarinen, n.d.)

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Occupational Therapy for Children with Severe Visual Impairment

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A loss of vision impacts on the whole array of daily life activities for children with severe visual impairment (SVI), from learning an activity through observation to visually directed hand use (Roley, 1995). Children with SVI are typically delayed in developmental skills across domains and they have to learn about the world in a different way. For this group of children, aim of occupational therapy is to maximize their potential for functional behaviours through the use of other sensory channels such as auditory and tactile senses to compensate for their visual loss.

Infant Stage

In infant stage, early sensory stimulation is essential. Children with SVI cannot see the approach of people and objects and can exhibit a very defensive reaction to touch i.e. tactile defensiveness. They will not want to use their hands to explore the world, perform fine motor tasks, as well as perform self care activities. Beside the aversive response to touch, children with SVI may also show fear of movement through space (Snow-Russel, 2001). They avoid dynamic movements such as crawling and walking. Owing to these sensory defensiveness, they are often extremely resistant to change; and are setting up a cycle reinforcing resistance to new stimuli. Therefore, they are easily being misinterpreted as emotional unstable or fussy children by their parents or caretakers. In order to improve their over-reaction to touch and movement, therapist uses specific handling techniques such as firm stroking, light pushing and pulling on the joints. Besides, children with SVI are encouraged to touch different textures through playing with sand, rice, shaving cream, etc. They also need to practice more movement in space such as rolling, pivoting, crawling, walking, jumping, etc. Active movements initiated by the child are highly encouraged since it is more therapeutic than passive movement.

Children with SVI are deprived of adequate visual stimuli and, because of a lack of mobility, are often secondarily deprived of tactile, vestibular, kinaesthetic, and proprioceptive stimuli. Therefore, different kinds of sensory stimulation such as swinging, being brushed on different body parts and familiarizing with different environmental sound, are introduced as early as possible. In addition, early sensori-motor training helps to develop their awareness of body and environment, which is the foundation for their later development in fine motor, self care and mobility skills.

Hands with good tactile discrimination ability and finger dexterity are crucial for children with SVI in exploration and concept development. Children with SVI manipulate objects to detect their form and shape in order to build up concept of daily objects. Better hand function may lead to higher competence in their daily living skills as they grow up. Thus, training on the basic hand function such as auditory-guided and tactile-guided reach out, precise grasp and release, mid-line orientation and bilateral hand skills is emphasized in their early ages.

In this infant stage, parent support and education on understanding the child's problem and also on handling skills is essential. Occupational therapist will give advice on handling techniques in order to extend the child's sensori-motor exploration within home setting. The parents need to learn how to provide adequate sensory stimulation to the child in daily tasks e.g. dressing and bathing time, as well as how to position the child in different activities. Since play is one of the main treatment media for children, another important advice for parents is on the suitable types of toys for training children with SVI. Concerning their visual deficit, it is important to consider the texture, size, shape, weight and auditory effect of a toy when choosing it as a treatment media. In addition, advice on home adaptation such as space arrangement, adaptation of feeding utensils etc, is essential.

Pre-school Stage

In the infant stage, training emphasis is on building up the foundation skills; while in the pre-school stage, the emphasis is on some advanced skills in order to prepare for later learning. For example, in preparation for learning Braille, children with SVI need to develop good tactile discrimination and object recognition skills. After they attain the basic tactile discrimination skills, the training will focus on the efficiency aspect i.e. the child has to perform not only accurate but also fast tactile discrimination abilities. In addition, this group of children needs to be equipped with advanced fine motor skills and adequate grip strength so as to enhance their independence in self care skills. Activities such as orienting and placing small pegs on pegboard, playing therapeutic putty, heavy-pulling and pushing games are included in the treatment sessions. Furthermore, on top of the basic bilateral coordination skills, children with SVI need to learn to use one hand as a reference point for the other hand to search in order to locate and search object efficiently.

The training of self care skills needs to begin early. It is difficult to motivate older children to look after themselves if they have got used to having everything done for them. Thus, basic self care skills such as eating and dressing skills are introduced in the early stage. Feeding aids can improve the efficiency of the feeding process. For example, the use of suction bowl can prevent knocking off of the bowl while scooping. Some special techniques such as holding his cup with one finger inside in order to know where to stop pouring water are being taught.

For enhancing their independence in mobility skills, it is important to develop good spatial concept, especially the laterality and directionality skills. They must know the concepts of left/right, up/down, in/out, etc in order to move freely within the environment. They have to develop the awareness that things outside the body have sides and must be able to measure distance and direction. They have to learn how to form a mental map of the environment and how to locate their working table, classroom, toilet, etc.

School Age

When children with SVI enter school age, they spend most of their time in school and also in the activities of daily living. Therefore, besides providing direct intervention, occupational therapist has an important role in liaison with school educators for promoting their understanding of the child's problem, as well as providing recommendation on environmental adaptation. It is important to modify the tactile and auditory environment around the child in order to compensate for his loss of visual feedback. For example, the use of tactile identifiers on different doors and floors, and the building of sound feedback into some items can facilitate children with SVI in searching and orientating to the school environment.

Another important training objective is to enhance their independence in activities of daily living skills, which can provide the children a sense of control and mastery and is essential in building up their self-confidence. The activities of daily living skills include self care skills such as dressing, grooming, bathing, and also the basic community living skills such as living district orientation, transfer between home and school, simple shopping, use of public facilities like pedestrian traffic light, etc. For the community living skills, occupational therapist has to work closely with the family and also the school in order to provide a tailor-made training program for the children.

Conclusion

Working with children with SVI offers great challenges to all of us. The visual deficit affect the child's development in all aspects and thus a multi-disciplinary team approach is highly recommended. Occupational therapist, as one of the members in the team for managing children with SVI, works closely with other team members in order to provide a holistic rehabilitation plan for this group of children. Although children with SVI cannot perceive the world through their eyes, they have great potential to learn and live independently with the use of other senses, which is also the ultimate goal of our intervention.

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A Quest to the Visual Function of Children with Cortical Visual Impairment and Severe Multiple Disabilities

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Prologue

CKY (Figure 1) was a quiet girl of two years of age with amiable temperament. The bare muscle strength in her neck afforded to turn the head to the side only when little CKY consigned to a recumbent position. Lying contently and yet motionlessly, CKY showed no excitement to any colourful merchandize which was no doubt adored by the young souls of her age. The calm and non-inquisitive expression was occasionally flavoured by a flash of smile in return to the long acquainted voice of her parents or a novel sound of a clinking chime. The easy-going temperament of CKY showed no intention to resist the highly skilled professional handling of the therapists in all positions and yet the tremendous support given as a result of the weakness in the neck and trunk together with the so-called spasticity in the limbs assigned her an official motor status as "a lyer". The little response which the rehabilitation and medical professionals endeavoured to elicit gave no better evidence than to prove her intellectual capacity being severely challenged. To complicate her identity further, her vision was in doubt and a diagnosis of cortical visual impairment aroused an uncertainty on her prognosis in all areas of development. Her health gave her no better position either; she had epilepsy which had not been fully under control yet and her swallowing pattern aroused suspicion of aspiration. Feeding had to be done with great care and time. To compete with the clock, the anticonvulsive drugs left her short spouts of arousal during the day. Despite the multiple challenges that lied in front of her, CKY was sure to have full support and care of her two dedicated parents.

Figure 1: Weakness in the neck and trunk makes sitting up very difficult for CKY.

The picture of CKY raised a question of where the focus of training should be if there was still room for it. Moreover, in face of her multiple disabilities a number of which related to her daily care, would the training of the visual function still be significant?

LWK (Figure 2), a boy of about the same age as CKY and sharing a similar diagnosis of spastic type of cerebral palsy affecting four limbs and with cortical visual impairment, presented initially

Figure 2: LWK often goes into extensor thrust on exertion.

himself with a totally different temperament from CKY. He was better off with a stronger neck and trunk and yet any attempts to interfere with his consoling and safe position in his mother's arms were met with strong protest of prolonged outcry of anger and pouring of tear. The colourful physical world did not seem to appeal to his visual appetite. Occasionally, the turning on of the neon light in the room appeared to wake his soul. He enjoyed a better health than CKY. However, the easily agitated state refrained him from any possible channels to reach his mind and body.

The picture of LWK posed another challenge to the staff at the forefront; how to open up learning opportunities for him.

The Group

CKY and LWK were two of the children in a group of six who shared similar challenges to the physical, cognitive, communicative and social development to a greater or lesser extent. The group was formed in September 2002 at the Jockey Club Conductive Learning Centre of the Spastics Association of Hong Kong because they all shared the common problem of cortical visual impairment (CVI) in addition to their severe physical and cognitive impairment.

The questions posed to the situation of CKY and LWK were the pivotal points of discussion on the long- and short-term management of the group. In spite of the multiple challenges and the dim prognosis lying ahead of this group of children, the team of staff including the childcare worker, the physiotherapist, the occupational therapist, the speech therapist and the nurse decided not to undermine the focus on creating maximal amount of learning opportunities for these children based on the fundamental belief that every child can learn. The training for visual function is deliberately not to ignore for two reasons:

1. The more channels of input available to a child who is already multiple impaired, the more tools he/she will have to make sense of his/her physical and social world;

2. Unlike sound stimulus, the non-temporal nature of visual stimulation is thought to be more apt for facilitating spontaneous exploration of the physical world. This inquisitive ability is particularly crucial for opening up a child who is locked in by his/her physical and cognitive incompatibility for further development.

The behaviour of LWK is typical of children with multiple impairments who have been overprotected and have little experience to "work under demand". The situation has to be unwound without delay before the child falls into the habit of "learning not to learn".

The Programme

The multiple impairments of this group of children entail the expertise of multiple disciplines which are now often available in a special childcare centre. However, the crucial point is how to make the best use of this multiple inputs of expertise to maximize the learning opportunities for this group of children. The Centre adopts the principles and practices of Conductive Education and finds they are particularly relevant to children with multiple impairments in the following perspectives:

1. All the professional staff forms a transdisciplinary team to tap on one another's expertise to plan and implement a holistic learning programme which integrates the physical, cognitive, communicative and social elements as a whole. In this way, the interconnectedness of the different challenges is attended to and consistency in handling is achieved. Interruption for taking each child to different departments for training is avoided. In doing so, the children will have a stable background for learning. This is deemed essential and beneficial for children who have limited channels to appreciate this busy world.

2. Learning in context is emphasized with a well-planned daily routine. With consistency and repetition, the children can gradually anticipate and comprehend what is going on around them. Familiarity breeds response. This is especially effective in establishing learning intention for children like LWK. For the parents, the learning programme is made so concrete and directly related to daily living that they can easily transfer the skills at home and create more opportunities for their children to practice.

3. Learning is made conscious with well-structured stepwise task (called task-series) linking up with pre-planned verbalization of the actions in rhymes or songs (called rhythmical intention). We find the rhymes and songs help the children to associate the movements required for the task as well as have a calming effect on them. The breaking up of the tasks into small steps allows a more organised introduction of stimuli, which fits well to the slow processing time of children with multiple impairments.

It is not the intention of this paper to discuss Conductive Education at length but to base on this background to illustrate how we have explored ways to stimulate the visual functions of this severely impaired group of children.

The Room and Teaching Material

All activities of daily living and learning programmes are carried out in the same classroom throughout the day – a stable and familiar background – which is favourable for creating a sense of security for children with CVI.

A simple plain background in the classroom is set up to reduce visual loading and provide contrasting background for targeted visual foreground (Figure 3).

Teaching material used in the learning programmes is carefully designed to arouse visual response (Figures 4 and 5).

The daily routine

Every moment is a learning opportunity – learning starts once the children enter the classroom the first thing in the morning (Figure 6).

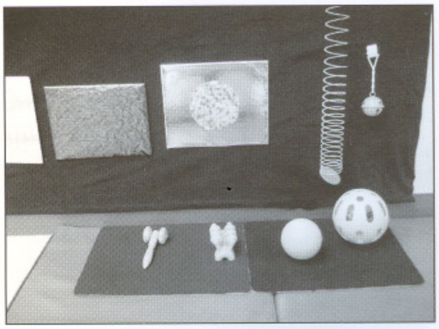

Figure 3: A corner in the classroom.

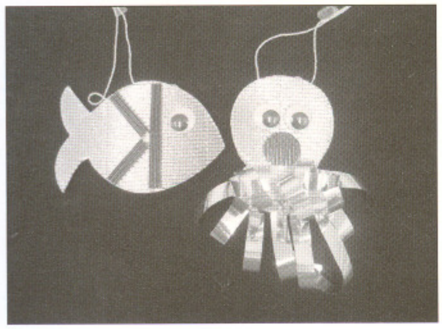

Figure 4: Teaching aids made with shiny material and contrasting colour to the background.

Figure 5: Washing bowl with a dark bottom to alert child's visual attention to her hand.

Figure 6: During waiting time, LWK was positioned to look at a fluorescence spring.

Meal time routine is a favourable time to create situations for using visual function in context. For example, a child learns to recognize his own eating utensils (Figure 7, 8 and 9).

Lesson time

Opportunities to use visual function are incorporated into different lessons and task series (Figures 10 and 11).

The Collaboration with Expertise

It is an innovative attempt for the Centre to systematically investigate the training of visual functions incorporating into the existing Conductive Education programme for these severely impaired children. We know the challenge is not easy to surmount. Thanks to the generous help of the experts in the field of visual impairment, the Visual Impairment Team of Child Assessment Service and the training team of the Ebenezer's Early Intervention Programme for the Visually Impaired Children. Here, we would like to share our experience in the collaboration with these experts in light of coordinating services for the best benefit of the children and their families.

The aim of the collaboration is to pull in existing resources to help these children and yet without confusing and exhausting the parents. To start with, we have the staff of the three parties meeting the parents together to explain the mode of collaboration, clarify any doubts from the parents and seek their consent for sharing their children's information among us. The assessment and main core of training is carried out at the Jockey Club Conductive Learning Centre of the Spastics

Figure 7: Presenting a white plate in front of a black board for contrast.

Figure 8: A red spoon on a white plate shows great contrast.

Figure 9: LWK looked at adult filling his plate with anticipation.

Figure 10: A visual board with contrasting pattern was presented after LWK responded to his name in roll call.

Figure 11: Playing with yellow paint in a black bowl incorporated tactile stimulation with visual stimulation for CKY.

Table 1: Visual function of CKY and LWK assessed by Child Assessment Service

	Visual performance in first assessment	Visual performance in second assessment
CKY	Light perception only (January, 2003)	Occasional fixation on large objects, faces, or movement (July, 2003)
LWK	Light perception only (November, 2003)	Visual acuity around 6/24 at 50 cm distance (July, 2003)

Figure 12: With the help of a gaiter and some manual assistance, CKY initiated some raking movement in her hand while looking at a piece of shiny paper.

Figure 13: LWY learning to walk with a ladder frame with centre staff, detached from his mother.

Association of Hong Kong as the Centre is the most familiar place for the children. The Visual Impairment Team of Child Assessment Service assesses the children twice a year. The results are immediately shared and discussed with the Centre staff and the Ebenezer's team. We find the immediate feedback from the assessment is particularly useful as there will be no delay for us to use the information for upgrading or adjusting our training programmes. The Ebenezer's team works closely with us on the training side. We set common goals for visual training. The team comes once monthly to join our lesson time in action. Feedback to the parents is followed by a discussion between the two team members on teaching techniques and programme refining. Some of the teaching material illustrated above is the product of such collaboration. The Ebenezer's team also gives home training and the programme is in line with the training at the Centre. Parents feel free to clarify any doubts with both teams of staff and find the consistency in our approaches.

The Preliminary Results

The collaboration began in October 2002. There were pleasing results concerning the visual function of the children particularly CKY whose parents were diligently carrying training systematically over at home. She has also shown signs of slight voluntary movement of the hand in attempt for reaching within near distance (Figure 12). LWK has also shown improvement in his visual function as well as his learning behaviour (Figure 13 and Table 1).

Epilogue

The small progress that these children have made is still far from changing their functional level drastically. The journey to achieve any significant results is long. However, in the process, the children have experienced the joy of learning which is part of their right no matter how severely involved they are.

Announcement and News

The Society has entered her Tenth Anniversary this year. We will organize a series of celebrations for this important milestone of our development.

We are very pleased to announce that Dr Robert Armstrong, the Vice President of the American Academy of Cerebral Palsy and Developmental Medicine, has accepted our invitation to be our Course Director at the Tenth Anniversary Annual Scientific Meeting. The theme of this year's meeting will be Rehabilitation and Developmental Paediatrics. It is scheduled for 19 – 22 November 2004. Please mark your diary and watch for future announcements. We will also open a free paper presentation during this ASM. It will be a good opportunity to share your experience and expertise.

Upcoming Meetings

Bimonthly Scientific Meeting

2 April 2004 (8 pm)

MG Lecture Theatre, Queen Elizabeth Hospital

Myasthenia Gravis
by Dr KH Chan, Department of Medicine, Queen Mary Hospital

Neuro-developmental Conference on Vision and Visual Impairment

19 May 2004 (8 pm)

MG Lecture Theatre, Queen Elizabeth Hospital

The 7th Annual Meeting of the Infantile Seizure Society: International Symposium on Neuronal Migration Disorders and Childhood Epilepsies-

Date	:	16 – 17 April 2004
Venue	:	Conference Hall, Tokyo Women's Medical University, Tokyo, Japan
Website	:	www.field.gr.jp/ISS

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8th Asian-Oceanian Congress of Child Neurology

7 – 10 October 2004

Hotel Taj Palace, New Delhi, India
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