Paediatric Ophthalmology
(in 120 minutes!)
FRACP Trainees – 4 July 2016
James Elder
Department of Ophthalmology, RCH
Outline of Lecture

• Examination Techniques
  – Vision
  – Ocular motility & Binocular Vision
  – Neuro-ophthalmology
  – Assessment of globe

• Specific Conditions
What is vision?

• Simple
  – Acuity, colour, contrast, field, depth perception, motion

• Complex
  – Extracting meaning
Examination Techniques
Assessment

• Objective

• Subjective
Normal Visual Expectations

• Birth

  – Brief fixation on
    • face
    • object
    • light
Normal Visual Expectations

• Six weeks
  – Maintain fixation on face, object or light and follow through 90 degrees
  – Smiling and visual responsive (especially to a face)
Normal Visual Expectations

• Six months
  – Reach for small object
  – Actively follow object
Normal Visual Expectations

• One year

  – Reach for tiny objects (eg 100’s & 1000’s)
Normal Visual Expectations

• 2 - 3 years
  – Picture matching (Kay picture test)

• 3 - 4 years
  – Letter matching (Sheridan Gardiner test)

• 5 years
  – Snellen acuity
  – logMAR tests
How to assess an infant’s vision

• History
  – Ask the parents
  – Family history of eye problems

• Observation
  – Watch child’s visual behaviour

• Tests
  – Elicit visual behaviour
Visual Acuity Tests
When to refer?

- Suspect poor or sub-normal vision
- Visual acuity of 6/12 or worse
Special tests of visual function

• Colour vision
• Visual field
• Stereoacuity
• Contrast sensitivity
Colour Vision
Visual Field

Perimetry

FRACP 2016
Visual Field

Confrontation
Stereoacuity
Ocular Motility

• \textbf{Duction} = movement of one eye

• \textbf{Version} = movement of both eyes

• \textbf{Duction} \neq \textbf{Version}
# Ocular Motility

## Ductions
- Abduction
- Adduction
- Sursumduction (Elevation)
- Infraduction (Depression)
- (Cycloduction)

## Versions
- Dextroversion
- Laevoversion
- Elevation
- Depression
- Convergence
- Divergence
- Cycloversion
Ocular Motility
Classification of squint - simplified
Binocular Vision

• **Aim:** stereoscopic depth perception

• **Downsides:**
  – Diplopia
  – Amblyopia
Binocular Vision

• Phoria vs Tropia

• Phoria = latent misalignment of eyes

• Tropia = manifest misalignment of eyes
  = more significant problem
Detection of Squint

- Observation
- Cover test
- Eye movements
Detection of Squint

- Observation
- Cover test
- Eye movements
Detection of Squint

- Observation

- Cover test

- Eye movements
Detection of Squint

- Observation
- Cover test
- Eye movements
Detection of Squint

• Observation

• Cover test

• Eye movements
Cover test
Detection of Squint

• Observation

• Cover test

• Eye movements
Additional examination

• Visual acuity

• Red reflex (children)

• General examination
Red reflex
Neuro-ophthalmology

- Pupil reactions
- Optic nerve function
- Examination of optic disc
- Visual field defects
- Nystagmus & odd eye movements
Neuro-ophthalmology

• Pupil reactions
  – Direct
  – Consensual
  – Near reflex
  – Relative afferent defect

  – Beware of neonatal & pharmacological miosis
Neuro-ophthalmology
Anatomy of Pupil Response

Fig 48. Diagram of the pathways taken by melanopsin ganglion cells to the brain. The pathway is to the suprachiasmatic nucleus (SCN) via the retinohypothalamic tract (blue) and on to the pituitary to regulate melatonin release. This circuit drives the circadian oscillator. A second pathway is through the lateral geniculate nucleus to the olivary pretectal nucleus (OPN) and to the Edinger-Westphal nucleus (EW) for control of the pupillary light reflex (light blue). After Berson, 2003.
Neuro-ophthalmology

• RAPD = relative afferent pupil defect

• Sensitive measure of retinal and/or optic nerve dysfunction

• Compares afferent input to pupillary response of the two eyes
Neuro-ophthalmology

• Optic nerve function
  – RAPD
  – Colour vision
  – Red desaturation
  – Visual acuity
  – Visual field
Examination of Optic Nerve

- Direct ophthalmoscope
- Dilated pupil helps enormously (tropicamide 1% very safe)
- Dim room lighting
- Child fixes on distant target
- Approach 10-15° from visual axis
- Your right eye for child’s right eye, etc
Normal Optic Disc
Papilloedema
Optic disc hypoplasia
Optic Disc Drusen
Optic Atrophy
Neuro-ophthalmology
Visual field defects

• Horizontal cut off think optic nerve
Neuro-ophthalmology
Visual field defects

• Vertical cut off think chiasmal & retro-chiasmal
Confrontation field
Neuro-ophthalmology

- Nystagmus & odd eye movements
- Will cover later
Globe

- Detection of
  - Extra bits
  - Cloudy bits
  - Missing bits

- Most can be done with simple light source (direct ophthalmoscope)
Globe

- Red reflex
  - Direct ophthalmoscope
  - 0.5 – 1 m from child
Strabismus
Childhood squint - comitant
Adult squint - incomitant
Consequences of squint - I

Children

- Amblyopia

- Loss of binocular depth perception

Adults

- Diplopa
Causes of squint - children

- Primary failure of binocular control
- Abnormality of convergence or divergence
- Refractive error
- Loss of vision in one eye
- Genetic tendency
Accommodative esotropia
Consequences of squint - II

• In both adults & children the head may be turned to maximise binocular function (abnormal or compensatory head posture)
Management of squint

- Confirm presence of squint
- Measure vision (including stereoacuity)
- Document diplopia
- Measure deviation
- Measure refractive error
- Examine for other disease (ocular & non-ocular)
- Determine need for special investigations
Management of squint

• Prescribe glasses
• Treatment of amblyopia
• Surgery
Aims of treatment

- Treat any associated amblyopia
- Minimise/overcome any diplopia
- Maximise binocular depth perception
- Improve binocular visual field (convergent squints only)
- Improve cosmesis
When to refer

• Squint definitely present
• Squint cannot be excluded
• Large angle squint before 6 months of age
• Any squint after 6 months (even if intermittent)
• Urgent referral if red reflex is abnormal
Watery Eyes
Watery (& Sticky Eyes)
Watery eyes

- Epiphoria is the result of
  - Excess tear production
  - Deficient tear drainage
Corneal foreign body
Corneal abrasion
Subtarsal foreign body
An approach to watery eyes in children
Sticky Eyes & Infections
Sticky eyes

- Discharge consists of mucus & inflammatory cells +/- pathogens

- Does not always indicate infection
Gonococcal conjunctivitis
(Ophthalmia neonatorum)
Sac Abscess
Pre-septal cellulitis
Orbital cellulitis
Pre-septal vs orbital cellulitis
Pre-septal vs orbital cellulitis

Pre-septal
- Sick
- Swollen lids
- Normal vision
- Normal pupil reaction
- Normal eye position
- Normal eye movements

Orbital
- Very sick
- Lids red +/- swollen
- Vision may be down
- May have RAPD
- Proptosis
- Restricted eye movements +/- pain
Trauma
Trauma to child’s eye

• Physical
  – Blunt
  – Sharp/penetrating

• Chemical

• Electromagnetic
  – Thermal
  – Radiotherapy
  – Light
Hyphema - blunt trauma
Blow-out fracture - blunt trauma
Penetrating injuries
Chemical trauma!
Non-Accidental Injury
What causes haemorrhage?

• Loss of integrity of vascular system!
• Disruption of vessel
  – Cut or tear vessel
  – Burst /rupture vessel
• Failure of clotting mechanisms
• Abnormal vessel fragility
Potential causes of retinal haemorrhage in infants

- Direct trauma to globe
- Clotting abnormalities
- Transmitted pressure
Potential causes of retinal haemorrhage in infants

- Leukaemia
- Haemorrhagic disease of the newborn
- Sickle Cell retinopathy
- ECMO treatment
- Retinopathy of prematurity
  - As part of disease (frequent)
  - As result of handling with neck flexion & extension (uncertain)
  - As the result of Retcam examination (rare)
- Galactosemia
- Henoch-Schonlein purpura
- Thrombocytopaenic purpura
- Maternal ingestion of Cocaine
- Meningitis
- Intracranial vascular malformation
- Optic disc drusen
- Retinal astrocytoma in tuberous sclerosis
- X-linked retinoschisis
- Intraocular surgery
- Severe hypertension
- Galactosemia
- Homocystinuria
- Glutaric aciduria*
- Osteogenesis Imperfecta
- Osteoporosis- pseudoglioma syndrome
- Incontinentia Pigmenti
- Central retinal vein occlusion
- Infections
- Cytomegalovirus
- Herpes simplex type 6
- Herpes simplex type 1
- Malaria
- Meningococcal septicaemia*
- Streptococcus pyogenes meningitis*
- Strept. Pneumoniae meningitis*
- Congenital toxoplasmosis
- Fibromuscular dysplasia
- Terson’s syndrome**
- Asphyxia
Potential causes of retinal haemorrhage in infants with abusive head trauma

- Direct concussive force applied to the eye resulting in vessels disruption (this mechanism may be a subset of the vitreo-retinal traction mechanism below)
- Increased venous pressure transmitted from intracranial cavity or central venous drainage in the chest
- Increased intracranial pressure resulting in localized venous obstruction, either within the distal optic nerve or at the level of the optic disc
- Direct spread of intracranial bleeding along the optic nerve (subdural or subarachnoid spaces) into the eye
- Vitreo-retinal traction leading to rupture of retinal vessels and splitting or elevation of the retina and rarely retinal dialysis or detachment.
Finite element analysis

Globe deformation with vitreous traction
Globe deformation with vitreous traction
Anisocoria

(Unequal Pupils)
Unequal Pupils

- Simple anisocoria
- Pharmacological
- Third nerve palsy
- Horner’s syndrome
Approach to Anisocoria

Is there more anisocoria in darkness or in light?

More anisocoria in light than in darkness

Check light reaction

More anisocoria in darkness than in light

Good light reaction in both eyes

PHARMACOLOGICAL MIOSIS

Poor reaction

Poor light reaction in one eye

Examine iris sphincter at slit lamp

Sector palsy of iris sphincter
Near-light dissociation

Test for cholinergic supersensitivity with pilocarpine 0.1%

Sphincter is supersensitive

Check motility

Impaired light reaction

Sphincter is not supersensitive

ADIE TONIC PUPIL

Iris transilluminates/pupil margin torn/other mechanical disruption

MECHANICAL IRIS DYSFUNCTION

Yes

No

Test for anticholinergic blockade with pilocarpine 1%

No pupil constriction/Full motility

PHARMACOLOGICAL MYDRIASIS

Pupil constriction/Limitation of motion

CRANIAL NERVE III PALSY

No dilation

Dilation

1% Hydroxyamphetamine test

Check associated signs and symptoms

Dilation

No dilation

HORNER SYNDROME

Cocaine test 2–10%

Dilation lag of smaller pupil

No dilation lag

SIMPLE ANISOCORIA

Dilation

No dilation

MYDRISTIC

More anisocoria in darkness than in light

Look for dilation lag of the smaller pupil

No dilation lag

Cocaine test 2–10%

Dilation

No dilation

HORNER SYNDROME

1% Hydroxyamphetamine test

Check associated signs and symptoms

Dilation

No dilation

Pre-ganglionic or central Horner

Post-ganglionic Horner

MECHANICAL IRIS DYSFUNCTION
Horner Syndrome – Clinical Features

- Anisocoria (miosis)
- Ptosis (upper & lower eyelids)
- Anhidrosis
- Conjunctival hyperaemia (transient)
- Dilation lag (Anisocoria @ 5 sec > @ 15 sec)
- Iris heterochromia
- Harlequin sign (reduced facial flushing)
Anatomy of the oculosympathetic pathway

- **1st order neurons**
  - Hypothalmus to lower cervical/upper thoracic cord

- **2nd order neurons**
  - Cord to superior cervical ganglion

- **3rd order neurons**
  - SCG to eye

Clinical Assessment Confounders

- Essential (physiological) anisocoria
- Unrelated ptosis
- Local ocular causes of miotic pupil
Diagnostic confirmation

- Cocaine
- (Apraclonidine)
Mechanism of Action of Diagnostic Agents

- NE = norepinephrine
- a = apraclonidine
- α1 = alpha 1 receptor
- α2 = alpha 2 receptor

Martin TJ
Current Neurology and Neuroscience Reports
2007, 7:397–406
Comparison of Agents

<table>
<thead>
<tr>
<th>Agent</th>
<th>Action</th>
<th>Observation</th>
<th>Use</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cocaine 4-10%</td>
<td>Blocks noradrenaline re-uptake</td>
<td>Miotic pupil does not dilate</td>
<td>Confirms Diagnosis</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Gold Standard</td>
</tr>
<tr>
<td>Apraclonidine 0.5-1% (α2&gt;&gt; α1 agonist)</td>
<td>α1 agonist de-innervation hypersensitivity</td>
<td>Miotic pupil dilates more than normal (reversal of anisocoria)</td>
<td>Confirms Diagnosis</td>
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## Comparison of Agents

<table>
<thead>
<tr>
<th>Agent</th>
<th>Difficulties</th>
</tr>
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<tbody>
<tr>
<td>Cocaine 4-10%</td>
<td>Availability, Stability</td>
</tr>
<tr>
<td>Apraclonidine 0.5-1%</td>
<td>? Sedation and cardiovascular SE in young children</td>
</tr>
<tr>
<td>Hydroxyamphetamine 1%</td>
<td>Not commercially available, False +ve with trans-synaptic degeneration</td>
</tr>
</tbody>
</table>
Cocaine test
Possible causes of Horner’s syndrome (!)

- **First-order neurons**
  - Pathology involving the hypothalamus, brainstem, and spinal cord
  - Infarction
  - Haemorrhage
  - Neoplasm
  - Syringohydromyelia
- **Second-order neurons**
  - Bronchogenic carcinoma
  - Sympathetic schwannoma
  - Neuroblastic tumors
  - Enlarged thyroid
  - Neck and upper thoracic surgery or other trauma
  - Primary spinal nerve root tumours
  - Osteophytes

- **Second-order neurons (continued!!)**
  - Jugular venous ectasia
  - Subclavian artery aneurysm
  - Other neck lesions causing compression of the cervical sympathetic chain
- **Third-order neurons**
  - Internal carotid artery dissection
  - Fibromuscular dysplasia
  - Skull base lesions
  - Cluster or migraine headaches
  - Agenesis or arteritis of the internal carotid artery
  - Aneurysm of the petrous portion of the internal carotid artery
  - Cavernous sinus inflammation, infection, or neoplasm
  - Rarely, orbital pathology: trauma or neoplasm after surgery
When to investigate?

- **Acquired** - all require thorough investigation if no pre-existing diagnosis or history of iatrogenic injury
  
  - Clinical examination
  - Urinary catecholamine assay
  - Imaging head & neck & chest (MRI > CT)
  - ? MIBG scan (meta-iodobenzlguanidine)
When to investigate?

• “Congenital”

• Often clinical uncertainty about time of onset

• Question: How likely is an isolated Horner’s syndrome in an infant to be due to something sinister?
Pediatric Horner’s with no known diagnosis at presentation

(CHOP, Philadelphia 1993-2005)

<table>
<thead>
<tr>
<th></th>
<th>Number</th>
<th>Comment</th>
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<tbody>
<tr>
<td>Patients</td>
<td>28</td>
<td>Further 28 with known diagnosis</td>
</tr>
<tr>
<td>Age years</td>
<td>2</td>
<td>Range 0.12 - 8.8</td>
</tr>
<tr>
<td>Idiopathic</td>
<td>21</td>
<td>1/56 birth trauma</td>
</tr>
<tr>
<td>Reactive lymph node</td>
<td>1</td>
<td>? Age ? Acquired or not</td>
</tr>
<tr>
<td>Neuroblastoma</td>
<td>4</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>3 acquired Age 5 &amp; 6 mths &amp; 3 years</td>
</tr>
<tr>
<td></td>
<td></td>
<td>1 congenital Age 6 months</td>
</tr>
<tr>
<td>JXG</td>
<td>1</td>
<td>15 months - acquired</td>
</tr>
<tr>
<td>Ewings sarcoma</td>
<td>1</td>
<td>3 years - acquired</td>
</tr>
</tbody>
</table>
## Horner’s syndrome < 1 year

(Single practice 1978-1995 CS Hoyt)

<table>
<thead>
<tr>
<th>Condition</th>
<th>Number</th>
</tr>
</thead>
<tbody>
<tr>
<td>Idiopathic</td>
<td>16</td>
</tr>
<tr>
<td>Birth trauma</td>
<td>4</td>
</tr>
<tr>
<td>Known neuroblastoma</td>
<td>1</td>
</tr>
</tbody>
</table>
| Ganglioneuroma                 | 1      | Acquired Horner’s
| Neuroblastoma                 | 1      | History unclear
| Follow-up (years)             | Mean 9.3 & median 9 | No new diagnoses made

BJO 1998; 82:51-4  
George et al
Demonstrable Horner’s syndrome within the first 4 weeks of life confirmed by photographs or examination (HSC, Toronto & Wilmer, Baltimore – 10 year period)

<table>
<thead>
<tr>
<th></th>
<th>+ve obstetric history</th>
<th>-ve obstetric history</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number</td>
<td>16</td>
<td>15</td>
</tr>
<tr>
<td>No other diagnosis</td>
<td>15</td>
<td>10</td>
</tr>
<tr>
<td>Developmental delay</td>
<td>1</td>
<td>-</td>
</tr>
<tr>
<td>Congenital varicella</td>
<td>-</td>
<td>2</td>
</tr>
<tr>
<td>Neuroblastoma</td>
<td>-</td>
<td>1 (prior stridor &amp; neck mass)</td>
</tr>
<tr>
<td>Lymph node (? Reactive)</td>
<td>-</td>
<td>2</td>
</tr>
<tr>
<td>Follow-up (median/mean)</td>
<td>29 &amp; 35 months</td>
<td>58 &amp; 64 months</td>
</tr>
</tbody>
</table>
Neuroblastoma & the Eye

- Ophthalmic features 80/405
- 60/80 orbital secondary
- 14/80 Horner’s syndrome
- 9/14 presented with Horner’s (2.2% overall)
- 11/405 presented with opsoclonus

Investigation of Horner’s syndrome presenting < 1 year

• Acquired – INVESTIGATE THOROUGHLY

• Not obviously acquired
  – +ve obstetric trauma - ? Need to investigate
  – -ve obstetric trauma
    • Offer investigation: CXR & Urinary catecholamines
    • Clinical review
    • More extensive investigation (MRI, CT or MIBG) questionable
Leukocoria
White Pupil (= leukocoria)

- Anything within the eye that is pale will result in a white or absent reflex and not a red reflex

- Cataract
- Retinoblastoma
- Chorioretinal coloboma
Cataract

• If suspect cataract refer

• Many causes

• Most due to isolated lens protein gene mutations (AD > XL > AR)

• Investigate if other abnormalities
Retinoblastoma
Retinoblastoma

- Present with leukocoria, squint, reduced vision or family history
- Sporadic (one-off or new mutation)
- Inherited (autosomal dominant)
- Unilateral or bilateral
- Fatal if untreated, 98% survival with treatment
- Early diagnosis increases chance of preserving vision
Retinoblastoma
Two Hit Hypothesis
Retinoblastoma
Potential Treatments

• **Surgery**
  – Enucleation

• **Irradiation**
  – EBR, brachytherapy, proton beam

• **Chemotherapy**
  – Systemic, local (subconjunctival / intra-arterial / intravitreal)

• **Cryotherapy**
  – Transcleral (may require fornix incision)

• **Laser**
  – Thermotherapy / photocoagulation
Principles of Rb Treatment

• If small ⇒ local treatment

• If larger ⇒ reduce tumour mass & then local treatment (SALT)

• If massive ⇒ often remove eye (consider reduction especially if bilateral)

• May be different for bilateral disease
Chorioretinal Coloboma
Congenital Glaucoma
Big eyes
Primary congenital glaucoma

- Raised pressure in eye
- Stretches the young eye (<2 years)
- May damage the optic nerve and cornea
Primary congenital glaucoma

- Watery eye (epiphoria)
- Light sensitive (photophobia)
- Corneal haze or cloudiness (corneal oedema)
- Corneal enlargement (buphthalmos = “ox eye”)
Primary congenital glaucoma

Treatment

• Medical
  – Sometimes
  – Be aware of systemic side effects
    • β blockers
    • α agonists

• Surgical
  – Often
Eyelids
Droopy Eyelids
Lumpy lids
Wobbly Eyes
Wobbly Eyes (most is nystagmus)

• Motor
  – Congenital nystagmus
  – Variably reduced vision
  – Null point with compensatory head position

• Sensory
  – Anything that reduces vision significantly in the first 4 months
  – Albinism
  – Untreated bilateral cataract
  – Retinal abnormality
Nystagmus - Examples
Nystagmus - Examples
Not Nystagmus - Examples
Not Nystagmus - Examples
Not Nystagmus - Examples
Systemic Disease & Children’s Eyes

(This could involve a series of lectures for several months)
Systemic Disease & Children’s Eyes

- Down syndrome (any of the previously described eye problems)
- Prematurity (retinopathy of prematurity)
- Juvenile chronic arthritis (uveitis)
Retinopathy of Prematurity = ROP

- Disorder of retinal angiogenesis
- Seen in some premature infants
  - <500 g .. 30%
  - 500-749 g .. 10%
  - 750-999 g .. 3%
  - 1000-1250 g .. <1%
  - >1250 g .. “never”
International Classification of ROP (ICROP) 1983 (revised 2005)

• Enable uniform description & collaboration

• Key elements
  – Location
  – Extent
  – Stage
  – Severity
ROP Location & Extent
ROP Stage
ROP - Severity

Plus Disease

Pre-Plus Disease
ROP “disaster” = RLF
Aetiology of ROP

• Unclear

• Low birth weight

• Gestational age

• Supplementary oxygen
Management of ROP

• Screening to identify those at risk of progressing to retinal detachment

• Laser treatment for “type 1” ROP to prevent progression

• Long term follow-up to check for sequelae
CRYO ROP 1988

- RCT
- Threshold = severe stage three for 5 continuous of 8 non-continuous clock hours in zone 1 or 2 with plus disease

- Conclusion
- Treating ROP of value (!)
- Cryotherapy - 21.8% unfavourable
- No treatment - 43% unfavourable
ETROP 2003

- **Type 1 ROP**
  - Zone I, any stage ROP with plus disease
  - Zone I, stage 3 ROP with or without plus disease
  - Zone II, stage 2 or 3 ROP with plus disease

- **Type 2 ROP**
  - Zone I, stage 1 or 2 ROP without plus disease
  - Zone II, stage 3 ROP without plus disease
ROP Treatment

Type 1 (Stage III) ROP – before & after laser treatment
RCTs & ROP

- CRYO-ROP (1988) - treatment works
- Light-ROP (1997) - not caused by light
- STOP-ROP (2003) - extra O$_2$ does not turn off ROP
- ET-ROP (2003) - earlier treatment better
- STOP-ROP (2010) - ↑ O$_2$ sats of little help
- BEAT-ROP (2011) - anti-VEGF turns off bad ROP
- BOOST (2013) - aiming for lower target O$_2$ sats increases mortality & only reduces ROP slightly
ROP sequelae

- Refractive errors
- Strabismus
- Amblyopia
- Visual loss if retinal scarring or detachment
Juvenile Chronic Arthritis

- Some children with JCA develop anterior uveitis (iritis)

- Risk of cataract and glaucoma as complications

- Beware the PANAFY (= Pauciarticular, ANA positive, Female, Young)
Juvenile Chronic Arthritis
Juvenile Chronic Arthritis
Learning Difficulties & Vision

• Little convincing evidence of connection between vision defects & learning problems

• Parents often try unproven treatments (burden of guilt)

• Educational approach (with appropriate psychological testing) of greatest benefit
Support Services for Children with Reduced Vision

• Pre-school
  – Early childhood educators from Vision Australia

• School age
  – Visiting teacher service (EVAC Clinic)
  – Vision Australia School

• Vision Australia Low Vision Clinic
  – Low vision aides

• Guide Dogs Association
  – Mobility & occupational therapy