Looking at the Ends of the Strabismic Spectrum – Infantile Esotropia and Paresis/Palsy of EOM

Congenital vs. Infantile Esotropia

- Nixon, Helveston, et. al. study (0 of 1219)
- Archer, Sondi, von Noorden study (0 of 3324)
- PEDIG 2002-175 infantile eT (40% reported at birth by parents)
  - Trust parents observations? Scan pattern?
- Scheiman and Wick reviewed infantile esotropia - found 28-54% of all esotropia is infantile in origin, the LARGEST group!
- Treat this as an infant, child or adult?

Infantile vs. Accommodative

<table>
<thead>
<tr>
<th></th>
<th>Infantile</th>
<th>Accommodative</th>
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<tbody>
<tr>
<td>Onset</td>
<td>B-6 mo</td>
<td>&gt;6 mo to 7 yrs</td>
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<tr>
<td>Angle</td>
<td>25-60PD</td>
<td>10-40PD</td>
</tr>
<tr>
<td>Refraction</td>
<td>&lt;+3.00D</td>
<td>&lt;+3.00D</td>
</tr>
<tr>
<td>Amblyopia</td>
<td>uncommon</td>
<td>common</td>
</tr>
<tr>
<td>LN, MLN</td>
<td>common</td>
<td>uncommon</td>
</tr>
<tr>
<td>DVD, IOOA</td>
<td>common</td>
<td>uncommon</td>
</tr>
<tr>
<td>Motion Asym</td>
<td>common</td>
<td>uncommon</td>
</tr>
<tr>
<td>VOR</td>
<td>common</td>
<td>uncommon</td>
</tr>
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Causes of Infantile Esotropia

Eye Scan 2-3 months
**Eye Scan 4-6 months**

- The whole is greater than the sum of its parts.
  
  Gestalt Theory

**PEDIG Studies of Infantile Esotropia**

- Clinical Spectrum of Early-onset Esotropia
  - Amblyopia frequently develops? (increases post-surg?)
  - The Natural History of Infantile Esotropia
    During the First Six Months of Life
  - Early surgical Tx may lead to increased coarse stereo?
- Spontaneous Resolution of Early Esotropia
  - <20 wk, <40PD, int or variable-like to resolve
  - Constant >40PD after 10wk-not likely to resolve
- *CONCERN – abduction deficit, off axis eT

**Etiological Factors of Infantile eT**

- Abduction Deficit/Pseudoparesis
  - Why not recognized? What if it disappears later?
  - Atrophy vs. Contracture, Primary vs. Secondary actions
- Cross Fixation
  - Abducting eye should lead localization - West
- Motion Processing Asymmetries/Latent Nystagmus
  - Nasal to Temporal develops with stereopsis-Tychsen
- Suppression and DVD, IOOA
  - Dorsal Light Reflex - Brodsky
- Others – Poor vergence? Because of X fixation?

**Abduction Deficit – Underreported?**

- DEFINE: Paresis vs. Palsy vs. Pseudoparesis
- Bourron-Madignier, Hugonnier-Prevention and treatment of amblyopia in children under two.
  - Basic motor disorder is abduction deficit, if asymmetric-can develop amblyopia.
  - *95.4% demonstrated abduction deficit (63 of 65)

**What Leads to an Abduction Deficit?**

- RARE-bilateral sixth nerve palsy, uni-maybe
- Birth trauma
- Viral infection
- Vaccination – 2,4,6 mo 6 types, MMR-cases reported
- Impingement of CN 6 by petro-sphenoidal ligament?
- Maldevelopment of oculocentric direction – Should lead by abduction, cross fixate leads T-N basis
- Precipitated by ear, nose, throat infection-petrous infl.
- Other – CN3 myelinated prior to CN6
Asymmetric Motion Processing

- At birth T to N is present (subcortical)
- Monocular N to T begins 2-3 mo., est. 4 mo.
- Symmetry by 9 mo., cortical in origin
- Confusion using OKN as visual acuity tool?
- Visual experience needed for maturation
- Stereo parallels N-T development of OKN at 4-6 mo., *Critical consideration with abduction concerns!

- *Can be used as differential test for time of onset / type of esotropia
- Likely on a continuum (qualitative)
- Right and Left not necessarily symmetrical
- Evaluation – OKN, Pursuits, Motion VEP
- Basis for Latent Nystagmus, or also vest.?
- Key - evaluate MONOCULAR N-T !!!

Development of Motion Pathway

- Abduction deficit sets the stage
- Cross Fixation develops for localization
- Motion Processing initially T to N, Cross fixation does NOT allow N to T to develop
- Establishes infantile eT and decorrelation of sensory development (loss of initial stereopsis)
- Abduction then generally releases, not eT

Monocular Pursuit Asymmetry

Big Picture of Infantile eT

Traditional Medical Treatment

- Monocular Patch – what is goal?
- Surgery – now pushed earlier, vs. europeans
  - Why earlier?
- Botox
- Prisms – Birch and Stager study
- Non-Surgical Treatment – Rethy, Sarniquet-Badoche and others
Birch and Stager Prism Study

Compensatory prism with infantile esotropia
- Up to 4 mo. strabismics showed some stereo, equal to normals
- After 4 mo. the stereo still dropped off with compensatory prism in place vs. normals

Why?
- Ocular motor may not be symmetrical – development of atrophy / contracture
- Lack of proprioceptive support
- No change in abduction deficit or motion asymmetry

Why Doesn’t Prism Maintain Stereopsis?

Mistake of Strabology - Rethy

- Miss the possibility of prevention
- Believe in unknown causes
- Apply symptomatic tx, not causal therapy
- Does an Optometric approach cover these?

“Treatment should be directed toward what is best for the patient, not for the surgeon.”
Stefan Rethy, MD
Considerations in the Literature

- Spontaneous Recovery
- Amblyopia
- Stereopsis
- DVD, IOOA
- Surgery vs. Non-Surgical

Vanishing Infantile Esotropia
Clark and Noel Canadian J Ophthal 1982;17(3):100-2

Three cases of large angle infantile esotropia are presented. Over a minimum of 37 mo the angle decreased spontaneously to less than 10 prism diopters without surgery or the use of glasses.

Complications of infantile esotropia developed including: bilateral IOOA, latent nystagmus and dissociated vertical deviation.

Spontaneous Resolution of Infantile Esotropia

3 cases diagnosed with infantile esotropia prior to 6 months demonstrated spontaneous recovery of their condition. By age 34 to 59 months the esotropia had changed to exophoria, esophoria less than 4 PD or orthophoria.

These patients showed relatively poor stereoacuity, dissociated vertical deviation and overaction of the inferior oblique muscles.

Untreated essential infantile esotropia : factors affecting the development of amblyopia.
Calcutt and Murray, Eye 1998;12(2):167-72

Amblyopia following early surgical intervention in essential infantile esotropia is well documented(“35-72% if you include patching), but the risk is outweighed by the chance of obtaining some form of binocular vision.

Where access to ongoing therapy is not available, patients with essential infantile esotropia, free alternation and no anisometropia have a significant chance of retaining good visual acuity (86-100%) if surgery is delayed until visual adulthood.

Spontaneous Recovery

- PEDIG Study
- Smaller angles, more intermittent appear to recover easier
- Infants who do NOT develop vergence 3-4 mo, appear to develop infantile eT, is cross fixation a substitution for the development of vergence?
- Many pediatricians do not refer out and often suggest infants will grow out of it

What are benefits and side effects of early surgery for infantile esotropia?

<table>
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<tr>
<th>Condition</th>
<th>No Tx</th>
<th>Post-Surgery</th>
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<tbody>
<tr>
<td>Amblyopia</td>
<td>0-14%</td>
<td>41-72%</td>
</tr>
<tr>
<td>Stereopsis</td>
<td>Gross</td>
<td>Better quality-more?</td>
</tr>
<tr>
<td>Fusion</td>
<td>Monofixation</td>
<td>6-24mo no better</td>
</tr>
<tr>
<td>IOOA</td>
<td>15%</td>
<td>72-78%</td>
</tr>
<tr>
<td>DVD</td>
<td>2%</td>
<td>62-76%</td>
</tr>
<tr>
<td>Side effects</td>
<td>Development?</td>
<td>See listing, also anesthesia concerns</td>
</tr>
</tbody>
</table>
VonNoorden—Surgical Success

- Optimal – Before 2 years best
- Desirable
- Acceptable
- Non-Acceptable
- BEST OVERALL 1-3 is after 4 years


SUMMARY of Early Surgical Outcomes

- Ignores possible causes for infantile eT
- Amblyopia increases
- No more with Stereopsis, but better quality
- Fusion-no difference if surgery before 2 yo
- Increase in DVD/IOOA
- Side effects do occur
- Concerns about ADHD, LD, Cognitive and Behavioral concerns secondary to early anesthesia

The Cochrane Database of Systematic Reviews

Interventions for Infantile Esotropia (Review)
The Cochrane Library 2005 #4, Repeated in 2011

The main body of literature on interventions for IE are either retrospective studies or prospective cohort studies. It has not been possible through this review to resolve the controversies regarding the type of surgery, non-surgical intervention and age of intervention. There is clearly a need for good quality trials to improve the evidence base for the management of IE.

Optometric Literature on Tx of Infantile Esotropia

- London, Griffin, Mazer-13 mo surgery, 11 yo VT
- Christenson-post surgery, after one year old
- Forrest-8.5 mo phoric by 2yo, essentially motor therapy
- Maples-seen 6 mo, started tx at 18 mo, Topical Journal on Infants, OVD 37(3) 2006
- Data not found regarding abduction deficit, motion processing, IOOA or DVD, in any cases

Optometric Management

- Binasal and Sector Occlusion
  - Placement considerations
- Abduction Considerations
  - Abducting eye should lead localization, promote peripheral motor fusion, eye and gaze should match!
  - Stereopsis should follow, temporal considerations
- Motion Processing Considerations
  - Onset matches development of stereopsis
  - Perhaps key to gaze holding, LN and stereopsis

How Might Binasal Occlusion Work?

- Impedes Cross Fixation – KEY!
  - Decreases abduction deficit, the abducting eye should be leading lateral localization !!!
  - Promotes N-T motion processing !!!
- Prevents amblyopia, no need to suppress !
- Promotes peripheral awareness/fusion
- Prevents anomalous correspondence
- Promotes alternation of eyes, easier guidance activities and ADL
Why might binasal occlusion work?

- Vary width and effects upon Sensory-Motor
- Can place asymmetrically for promotion of alternation, amblyopia E/M
- This modifies visual field relationships, also known as penalization
- Modifies amount and quality of light approaching the eyes (suppression)

How might a binasal work?

Binasal Effects on Visual Input/Response

- Emphasis to localize monoc. with less confusion
- Nasal Retina- Localization  Motor Fusion  Decussate
- Temporal R.- Stereopsis  Non-Decussate  Cross at Corpus Callosum

Visual Field to Ocular Dominance Columns

Correlated and Decorrelated Activity:

Balanced Gaze, Motion, Fusion and Stereopsis
Motor essentially sets the stage for sensory development

Abduction Deficit - Therapy
- Pursuits
- Saccades (eye throwing?)
  - Monocular prism jumps
- OKN Cloth (motion), slow vestibular rotation
- VOR – alternate doll’s eye, PRN monocularly
- Vergence if fusing – in/out, lateral excursions
- Ron, et. al. study on oculomotor subsystem transfer
  *Binasal helps set the stage for biocular work

Motion Processing Therapy
- Binasal sets the stage for treatment
- Importance of treating abduction deficit
- Types – Pursuits, OKN, Vestibular input
- Thus you have an overlap of abduction and N-T motion processing therapy!
- ADL’s- feeding, diaper change, etc.

Vestibular Applications
- Parents including it daily, with fixation (mirror)
  - Consider prism with it therapeutically?
- Increase arousal – involved in therapy, postural control improved, suppression? (BO/BI ranges)
- *Slow away from abduction deficit for stretching
- *Faster to abduction deficit, use post rotary nystagmus to break abduction, synchronize with volitional looking/reaching
- Sue Barry speaks of NOVELTY in adult plasticity, what about infants?

How else can you facilitate the recovery of abduction deficit?
  *if you have a non-responder

Cranial Osteopathy
- Cranial pulse, rhythm
- Cranial molding and movement
- Petrosphenoidal ligament (ligament of Gruber)
- www.cranialacademy.com
- Upledger Institute – Chiropractic, others
Considerations of the Petrosphenoidal Ligament ala… Ligament of Gruber

- Origins are dorsum sellae(sphenoid) and petrous process(temporal bone)
- CN6 passes below it, before entering the cavernous sinus…
- But…2/3 are divided, half under and half above.
- Thus only 1/3 of cases would be possibly fully affected by the ligament…is this why we don’t see more abduction deficits?

Optometric Goals

- Decrease abduction deficits
- Decrease cross fixation
- Improve N-T motion processing
- Increase alternation
- Promote ipsi eye leading localization
- Emphasize peripheral fusion
- Do we “cure”, or manage the condition?

Case Examples

- Early onset, abduction releases early on
- Abduction won’t release, what else?
- Does infantile esotropia treatment include only infants? Or also adults who had it earlier?
- Adult infantile esotropia – well known example would be Susan Barry, PhD from “Fixing My Gaze”

Summary Overview-Infantile Esotropia

- Decrease abduction deficit and cross fixation
- Increase lateral tracking, motion processing
- Allows for peripheral fusion, decrease need for suppression, effects upon DVD and IOOA
- If you believe a surgical approach should be your first choice of treatment, what might be easier for a surgeon…treat an infantile esotrope with an abduction deficit or full range of movement? (hint: think consecutive exotropia)

Diagnostic and Therapeutic Considerations for EOM Paresis and Palsy

Curtis R. Baxstrom, MA, OD, FCBO, FNORA
Can We Treat a EOM Paresis/Palsy?

- Palsy-CN dysfunction or paralysis
- Paresis (partial palsy) - used for EOM, full or partial palsy?

What if OT and PT’s treated hemiplegia the same way that EOM paretic cases are often medically treated?

What does monocular patching provide the TBI patient?

Next Question: When should one begin treatment?

What Should We Do?

1. Support a discharge, nothing can be done visually.
2. Consider what he can and cannot do, what goals might we set?
3. Determine what we might do to help him achieve those goals (NOR and ADL).
4. Work with OT/Speech/PT in carrying out the plan.

Followup One Week Later

- Speech reports better visual attention to communication device (dynavox)
- Speech is last treatment of day, Speech reports he is no longer dozing off at end of day
- Overall better arousal and attention, no dozing off during my follow-up visit
  - Doll’s eye vs. VOR gain from lenses vs. Other?
- EOM – Typical 3-4 saccades during pursuit has reduced to 2 saccades, ROM and latency has also improved
- He is now EMERGING from his TBI!
Three Months Later

- Can move arms, hands and fingers, not walking, but can stand at table
- Easily using his dynavox
- Making one word utterances
- Can track freely in space. Was provided EOM paresis protocol.
- Near lenses beneficial
- Quality of Life has changed!!!
  - Not cured or back to pre-accident status

KEYS to Care

- EOM treatment is more than simply eye muscle exercises, includes vestibular input with others!
- By improving EOM control and adding vestibular input, the patient began to emerge from his TBI!
- KEY-Effects of Visual-Vestibular Input upon rehabilitation as a whole

Differential Diagnosis

- Origin – Developmental vs. Acquired
- Paresis vs. Paralysis
- Subcortical vs. Cortical pathways
- Innervational Deficiencies
- Mechanical Restrictions
- Direct Muscle Trauma

Special Considerations

- Effects upon EOM
  - Atrophy – paretic muscle
  - Contracture – opposing muscle
  - Muscle shortening – loss of cells
  - Vertical vs. Horizontal EOM
  - Spread of comitance-how fresh?

Testing for Paresis / Paralysis

- Monocular Range of Movement
  - Include Doll’s Eye – repeated?
  - Include binocular ranges if possible
- CT in all 9 Positions of Gaze
- Underaction and Overaction
- Parks 3 Step – isolate cyclovertical EOM
- Hess-Lancaster testing

Version Testing

- Hering’s Law of Equal Innervation
- Rate the movement on scale of 0-4
- Mark Overaction with +
- Mark Underaction with –
- Each number represents approximately a 25% change
**Muscle Action Scale**

<table>
<thead>
<tr>
<th>GRADE</th>
<th>% Over/Underaction</th>
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<tr>
<td>0</td>
<td>Normal Action</td>
</tr>
<tr>
<td>1</td>
<td>25%</td>
</tr>
<tr>
<td>2</td>
<td>50%</td>
</tr>
<tr>
<td>3</td>
<td>75%</td>
</tr>
<tr>
<td>4</td>
<td>100%</td>
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</tbody>
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*Look in 8 Positions of Gaze
*Consider Photodocumentation

**Parks Three Step**

- Which eye is hypertropic in primary gaze?
- Hypertropia increases in which gaze?
- Hypertropia increases with which head tilt?

**Considerations of Parks Three Step**

- Isolates single cyclovertical muscle
- Problems
  - What if multiple muscles?
  - Spreading of Comitance
    - Fresh vs. Old paretic cases
    - Paretic heals, antagonist has contracture
  - Doesn't differentiate paretic vs. fibrotic
  - Post-Operative findings may also be misleading

**Hess Lancaster Testing**

![Hess Lancaster Testing Diagram]

Patient wearing green and red glasses (provided) is given hand projector and directed to place green dot inside red circle. Relationship of dot to circle makes diagnosis possible.

**Hess Lancaster Testing - RLR**

![Hess Lancaster Testing - RLR Diagram]
Hess Lancaster - RLR,RSO

Considerations of Paresis and Palsy

- Patient needs -
  - Safety during mobility and ADL's
  - Recovery of function
- Patient, Rehab Team, OD goals
- Rehabilitation vs. Compensatory vs. Combined approaches

Traditional Treatment – Medical

- Monocular occlude – which one?
- Wait and see
- Botox to decrease contracture
  - Sometimes aligns patient, why not tx?
- Surgical considerations – 1 year

Another Look at the Medical View

- Why are EYES the only ones looked at from a “wait and see” approach?
- Benefits of unilateral full occlusion
  - No diplopia
- Side effects of unilateral full occlusion
  - Decreased binocular input, child adopts
  - Possible shift in visual midline/egocenter
  - Decreased visual field
  - Muscle effects – contracture vs. atrophy
  - Effects upon functional recovery

Optometric Considerations for Treatment

- Two Factors – balance the needs
- Range of Movement
- Diplopia Considerations
  - Occlusion – selective vs. full
  - Compensatory Prism
  - Guidance - Range of Diplopia

ROM Monocular Treatment

- 1-Pursuits
- 2-Saccades - Margolis Eye Throwing
- 3-OKN – www.BuyFabrics.com
- 4-VOR / Doll’s Eye – Vestibular input
  - Single vs. repetitive inputs
  - Post rotary nystagmus – bidirectional
- Additional aspects
  - Monocular prism jumps
  - Afterimage transfer
## Oculomotor Therapy Effects in Traumatic Brain Injured Patients

**Faster Rate of Improvement**
- Saccades: 4.5X
- Optokinetic: 3.0X
- Pursuit: 2.5X

*Also a higher level of improvement
*Some oculomotor subsystem transfer
See JBO article by Ciuffreda- EOM Rehab

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## S. Ron, et.al., continued

- Can training be transferred from one oculomotor system to another? Physiological and Pathological aspects of Eye Movements. Roucoux & Crommelinck eds. 1982:83-98
- Training Oculomotor Tracking Israel J of Medical Sciences 28:622-628, 1992

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## TBI Strabismus Overview

- Monocular - Paresis / Palsy
- Binocular – usually do not suppress, but can
  - Selective Occlusion
  - Prism – phasic and tonic aspects, visual and vestibular
  - Rehabilitation/Therapy – phasic and tonic aspects, visual, vestibular, motor

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## Selective Occlusion-Considerations

- Compensatory vs. Therapeutic (both?)
- Size
- Unilateral vs. Bilateral
- Nasal, Temporal, Superior, Inferior
- Form – sector, spot, full, etc.
- Opaque vs. Graded vs. Color

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## Example – L CN6 Paresis

- Look primary and L gaze-diplopia, R sees single
- L Temporal Sector-OT
  - Eliminates diplopia to L
  - No therapy on L abduction
- R Nasal Sector
  - Eliminates diplopia to L
  - Allows attempts to improve L abduction
  - Limitation of L gaze in mobility?
Therapeutic Prism

- Goal is to establish single vision and to facilitate removal over time
  - If unable to remove all, compensatory!
- KEY - Monocular ranges – enough?
- Is the binocular system established?
- Tonic (slow) vs. Phasic (fast) Vergence and Vestibular systems
- No Fusion- Sensory Fusion Disruption Syndrome

Vision Rehabilitation / Therapy

Three Uses of Vestibular Ocular Reflex

- Maintain postural control
- Kinetic/transitory contractions for maintenance of equilibrium and EOM during movement
- Maintain muscular tone
- Works with motion to control blur
- Can be used in VISION REHAB
- *ALSO – vestibular input and arousal

Vestibular Component-Types of Input

- Semicircular Canals – Phasic Input
  - Each canal to specific muscle
  - Rotational input
  - More likely used for esotropia
- Otoliths – Tonic Input
  - All EOM affected
  - Linear input
  - More likely used for exotropia
  - BUT…combine both for overall outcomes?
Therapeutic Prism Protocol

- Use Fresnel prism, minimum to fuse
- Changes in sensory, monocular vs. split
- Near vs. Far, Comitancy
- Weekly therapy vs. Periodic followup and modification of prism
- Every patient receives therapy including vestibular input
- GOAL-Reduce prism to zero, recheck fusional ranges, discharge, followup

Therapeutic Prism Protocol

- Therapeutic considerations-
  - Vestibular- may be key to modifying the Tonic Vergence System
  - Vestibular activities
    - Turn and clap
    - Turn and catch
    - Bean Bag activities
  - Supplement with traditional vergence therapy (Ludlam-jump duction)

Prism Considerations

- Benefits to increased peripheral field
- Benefits to visual midline and mobility
- Leads to treatment protocols for recovery vs. maladaptations
- Learned recovery of intermittent diplopia
- Overall rehabilitation process is supported-vision leading and guiding.

Case Presentations

Non-surgical treatment for esotropia secondary to Arnold-Chiari I malformation:
A case report.

Optometry 2009, 80, p.472-78.
(45eT to fusing 12 BI)
1 year diplopic prior to start
Surgical Treatment of ACM

- Posterior fossa decompression
- Suboccipital foraminotomy
- Duroplasty
- Decompressing the cranio-cervical junction
- Combination of above

Visual Findings -

- Case Hx: 3 year ago esotropia onset, 16 mo. post ACM surgery, esotropia still present, abduction recovered
- 45 prism diopter esotrope far and near
- -1.75 myope
- Ocular health otherwise unremarkable
- Started therapy at 20 months post ACM surgery

Visual Findings of 5 Month FU

- Eccentric Circles maintenance therapy has been done somewhat inconsistently
- Refractive Status R -.25, L -.75
- Good reserves in accommodation and convergence
- Saccades within normal as measured by DEM
- Visual hygiene and near plus was discussed

Three Year Followup
small increase in myopia

Seven Year Followup
post MVA

AI Transfer, RG Vectos

- After Image Transfer
- R/G Anaglyph, Tranaglyph – move head vs. tgt
  - Jump Recoveries
- Work to limits of capability
- Continued Eye Throwing and Post Rotary Vestibular Input
Summary Overview

- Start ASAP! Never too early!
- Visual needs of the patient in rehab
- Evaluate, set up treatment plan
- Recovery of Range of Movement
- Reestablish Binocularity – when to Rx Prism?
  - Occlusion
  - Compensatory Prism
- Support total rehabilitation approach

Visual Rehabilitation should be fuller and faster than the traditional unilateral patching regimen.

Thank You!

2016 NORA Clinical Skills Program
Level 1, 2 and Advanced
Austin, TX
January 15-17, 2016
*Next Annual Meeting Sept. 22-25, 2016