Is Albinism a Pain in the Neck?
The Pain in Albinism Pilot Study

Dr Shari Parker
St Vincent’s Hospital
Sydney AUSTRALIA

2EDA Valencia
April 5-6 2014

Images thanks to Positive Exposure and the amazing Rick Guidotti, Australia 2010
Today’s talk

• A bit about me
• Headache / neck pain in people with low vision
• The Pain in Albinism Study
• Rehabilitative approaches for pain in albinism
Sydney, Australia
Me and my sister
Dr Shari
University of NSW
16 to 18 October 2015
New Zealand
Massey University

ALBINISM
FELLOWSHIP
OF
AUSTRALIA
Background – The nature of the Problem

• Anecdotally, neck pain and headache common in PWA

• Research on pain in low vision?

• Research on pain in albinism?
Significance of the problem

Associations with chronic pain

• Poor Quality of life
• Psychological wellbeing
• Insomnia
• Functional, Social, Employment issues
• Mortality
• Cost – healthcare, society
Aims of Pain in Albinism Pilot Study

1. How common is pain in people with albinism (PWA)?

2. What impact does pain have?
   a) Severity
   b) Quality of life
   c) Health use
   d) Work / absenteeism
Aims Continued

3. How do people with albinism manage their pain?

4. What factors are associated with pain in PWA?
   - Photophobia
   - Lighting
   - Using vision aids
   - Daily activities eg sleep, exercise
Method

- Voluntary completion of paper questionnaire
- People attending albinism conferences in Australia, USA, Malaysia
- Being converted to electronic version
- Ethics approval St Vincent’s Hospital
Results
Disclaimer

• Low numbers
• Non random sample
• People with pain more likely to respond – greater interest
• Difficult to make generalisations
• ** indicates statistically significant difference
44 respondents
Gender

- Male 41%
- Female 59%
Education Level

- Elementary
- High School
- College
- Trade
- Bachelor
- Post graduate
Type of Albinism

- OCA: 80%
- OA: 0%
- Unsure: 10%
Pain
84% reported pain
Average pain severity 4.8/10
Worst pain Average 6.2/10
Frequency of episodes

- Constantly
- Daily
- Most days
- Few days
- Weekly
- Monthly
- < monthly
Impact of pain
Impact on usual activity

Average 3.6/10
# Health Care Utilisation

<table>
<thead>
<tr>
<th>Service</th>
<th>Episodes per year</th>
</tr>
</thead>
<tbody>
<tr>
<td>General Practitioner</td>
<td>0.51</td>
</tr>
<tr>
<td>Specialist</td>
<td>0.30</td>
</tr>
<tr>
<td>Admission</td>
<td>0.14</td>
</tr>
<tr>
<td>Allied Health</td>
<td>1.3</td>
</tr>
<tr>
<td>Alternative Practitioner</td>
<td>1.1</td>
</tr>
<tr>
<td>Imaging</td>
<td>0.19</td>
</tr>
</tbody>
</table>
Health Related Quality of Life

- Quality of life related to your health
- Individual’s perspective
- Physical, psychological, social
- Generic
- Condition specific
4. Health Related Quality of Life
Short Form 8 (SF8)

- 8 questions
- 8 domains
- Grouped into 2
- Physical
- Emotional
Quality of Life

Physical function

Role Physical

Bodily Pain

General Health

Vitality

Social Function

Role Emotional

Mental Health
Health Related Quality of Life SF8

- Two summary scores:
  - Physical Summary
  - Mental Summary
- 0 to 100
- 50 is population average
- High is good, low is bad
- Physical + Mental = HR QOL
General Health Item

• Overall, how would you rate your health during the past four weeks?
  Excellent       Very Good       Good
  Fair            Poor           Very Poor

• This item is a strong and significant predictor of:
  – Mortality
  – Use of health resources
  – Change in functional status
  – Recovery from episodes of ill health
Quality of life

- Physical
- Mental
- Total

**P = 0.01**

**p = 0.08**

**p = 0.003**
Quality of life

Pain | no pain

PF | **RP | **BP | **GH | **VT | SF | RE | MH

30 | 35 | 40 | 45 | 50 | 55 | 60 | 65
Pain predicting factors
Associated Factors

Predictive
• Photophobia
• Prolonged computer use
• Prolonged reading
• Flat desk
• Driving
• Fluoro and down lights
• Scoliosis

Protective
• Using sloped desk
• Using visual aids
• Walking
• Exercise
• Neck and back exercises
• Adequate sleep
• Indirect lighting
• Standard light globes
<table>
<thead>
<tr>
<th></th>
<th>Pain</th>
<th>No Pain</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Age</strong></td>
<td>38</td>
<td>43</td>
</tr>
<tr>
<td><strong>Gender</strong></td>
<td>33% male</td>
<td>86% male</td>
</tr>
<tr>
<td>Acuity</td>
<td>No significant difference</td>
<td></td>
</tr>
<tr>
<td>Smallest print</td>
<td>No significant difference</td>
<td></td>
</tr>
<tr>
<td><strong>Photophobia</strong></td>
<td>6.6 / 10</td>
<td>5.1 / 10</td>
</tr>
</tbody>
</table>

No significant difference for
- Living arrangements
- Marital status
- Education
- Employment
## Pain predicting factors

<table>
<thead>
<tr>
<th>Lighting</th>
<th>no significant difference between PWA with and without pain for different types of lights used</th>
</tr>
</thead>
<tbody>
<tr>
<td>Use of vision aids</td>
<td>no significant difference between PWA with and without pain</td>
</tr>
<tr>
<td>Scoliosis</td>
<td>16% of PWA with pain had scoliosis versus 0 in those without pain</td>
</tr>
</tbody>
</table>
Activities
<table>
<thead>
<tr>
<th>Activities</th>
<th>Pain</th>
<th>No pain</th>
</tr>
</thead>
<tbody>
<tr>
<td>Computer</td>
<td>3.5</td>
<td>1.4</td>
</tr>
<tr>
<td>Reading</td>
<td>0.9</td>
<td>0.9</td>
</tr>
<tr>
<td>Flat desk</td>
<td>2.4</td>
<td>0.8</td>
</tr>
<tr>
<td>Sloped desk</td>
<td>0.1</td>
<td>1.6</td>
</tr>
<tr>
<td>Walking</td>
<td>1.8</td>
<td>1.1</td>
</tr>
<tr>
<td>Exercising</td>
<td>0.3</td>
<td>1.1</td>
</tr>
<tr>
<td>Sleeping</td>
<td>6.6</td>
<td>6.7</td>
</tr>
</tbody>
</table>
Summary
Summary

• Pain is common in PWA 84%
• Neck pain and headache common
• Average pain 5/10
• 40% pain on most days of week
• Moderate impact on functioning
• Pain → poor health related quality of life, especially physical
Associated Factors

Risk factors
- Female
- Using computer
- Use flat desk
- Photophobia

Protective
- Male
- ? Getting older
- Sloped Desk
- Exercising
Discussion – why might PWA get pain?
And What can we do about it?
Why might people with albinism get pain?

1. “Asthenopia” eye strain
   Eye pain, headaches, neck pain, fatigue, blurred or double vision, burning or dry eyes, photophobia

2. Posture related
   ➔ Eye related
   ➔ Environment
   ➔ Personal factors
Why do PWA get pain?

<table>
<thead>
<tr>
<th>Eye related</th>
<th>Environment</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Photophobia</td>
<td>7. Computer</td>
</tr>
<tr>
<td>2. Scoliosis</td>
<td>8. Print size</td>
</tr>
<tr>
<td>3. Head tilt</td>
<td>9. Light</td>
</tr>
<tr>
<td>4. Strabismus</td>
<td>Personal factors</td>
</tr>
<tr>
<td>5. Refractive errors</td>
<td>10. Diet</td>
</tr>
<tr>
<td>6. Foveal hypoplasia</td>
<td>11. Stress</td>
</tr>
<tr>
<td></td>
<td>12. Fatigue</td>
</tr>
<tr>
<td></td>
<td>13. Psychological</td>
</tr>
</tbody>
</table>
Why might PWA get pain? What can be done about it?

1. Photophobia / light environment
   - Muscle overactivity / spasm
     - Constrictor pupillae (CNIII)
     - Corrugator
     - Obicularis oculi

✓ Sunglasses
✓ Hat – even when indoors
✓ Tinted contact lenses
✓ Avoid down and fluoro lights
✓ Indirect Light
Why might PWA get pain? What can be done about it?

2. Scoliosis

- 5* increase
- Scoliosis may be related to ↓balance
- Vision important in postural control and balance

✓ Scoliosis Screening

3. Head tilt – nystagmus null point

✓ Nystagmus null point surgery
Why might PWA get pain? What can be done about it?

4. Strabismus
   – Imbalance of extra-ocular muscles
   ✓ Corrective surgery

5. Refractive errors
   - Greater use of muscles for accommodation and convergence
   ✓ Correct refractive errors – glasses, ? Laser, insertion of intraocular lenses
Why might PWA get pain? What can we do about it?

6. Computer and print size

- Ciliary muscles (change lens shape) for viewing items at near distance (accommodation)
- Worse if uncorrected refractive errors / Eye muscle imbalance / strabismus
- Worse when reading on computers vs hard copy
Why might PWA get pain? What can we do about it?

Reduce asthenopia / Eye Strain

✓ Enlarge, audio, bring things closer
✓ Hard copy preferable to computer
✓ Regular breaks, 5 mins – focus in the distance
✓ Moisture – blink, artificial tears
Can we reduce Asthenopia with diet?
7. Diet?

• RCT placebo / active supplement 4 weeks
• 3 ingredients in supplement capsule

1. Fish oil (DHA, EPA) - Antioxidant, accumulates in retina, role in preventing MD
2. Bilberry Extract (authocyanins) – Antioxidant, helps restore rhodopsin
3. Lutein - Antioxidant accumulates in retina, especially macula

Treatment group – significantly lower scores on asthenopia questionnaire (stiff shoulder, dry eye, frustration, stuffy head), &↓ mental fatigue
Why might PWA get pain?  
What can we do about it? 

8. Poor macular development
   ➔ Close focal point
   ➔ Postural alterations to bring the eyes closer to the item of interest
   ➔ Hunching & poor posture

9. Stress / Fatigue / poor self esteem 
   ➔ poor posture
   – Study – people in erect vs hunched posture
   – Hunched posture ➔ helplessness, reduced persistence in tasks, greater sense of stress
   – Hunched posture associated with ↓MH
Poor Posture – loss of the neutral spine
ALIGNMENT – The Neutral Spine

- 3 Primary curves
- Cervical lordosis (in)
- Thoracic kyphosis (out)
- Lumbar Lordosis
- Should be subtle
ALIGNMENT – The neutral Spine

• Line up
  Ear hole
  Tip of shoulder
  Outer hip
  Mid knee
  Just in front of bone
MUSCLES

1. Movement - Sprint muscles

2. Scaffolding Marathon muscles
In the Neutral spine

• Marathon Muscles switched on
• Bones aligned
• Use 5 X less energy
• Slow down wear and tear
OUT OF BALANCE
OUT OF CONTROL
Do I have a Neutral Spine?

Wall test

Mirror test
Posture and Pain

• The Turtle Gorilla – neck pain and tension headaches. 1 inch = 10 pounds extra weight

• Factors associated with neck and upper back pain in school children
  – Prolonged sitting
  – Bending forward > 20 degrees
  – Depression and stress
“Bad” Posture
The Turtle Gorilla
Overactive muscles → Short and Tight → Fatigued

Build up lactic acid → Acute pain → Chronic pain

Wear and tear → Discs, joints → Arthritis, degen

Nerve compress’n → Organs – lungs, bowels → Poor self esteem
Benefits of Improving your Posture

😊 Less risk of chronic pain
😊 Better control of pain if present
😊 Improvement in self confidence – ↑ sense of own power and more likely to take action
😊 Perceived by others
  – Thinner and more attractive
  – More confident and successful
Improving Posture

✓ Object to the eyes
✓ Sloped desks
✓ Gadgets – CCTVs, Monoculars
✓ Make use of audio technology
✓ Ergonomics - Avoid forward postures
✓ Enlarge ENLARGE!
✓ Strengthening deep flexors (evidence)
✓ Range of motion, stretching
✓ Walk tall and proud – consider use of cane
Improving Posture

- Lose weight if necessary
- Exercise, especially in nature
- Firm mattress, single pillow, no tummy sleeping!
- Seek help for stress / anxiety / depression
- Improve self esteem
- Be Proud of who you are
- Relaxation, meditation
- Look on the “lighter” side of life 😊
- Correcting the “wall test”
What to do if you do have pain

Red flags

• Severe pain
• Doesn’t settle
• Sensory changes
• Weakness
• Bowel / bladder
If Problems do Occur

• Stretches, Hot pack, warm shower
• Simple analgesia
• Resume normal activity, don’t go to bed!
• TENS, traction, ultrasound, massage, mobilisation, manipulation......
• Prevention better than cure
• Seek medical attention if “red flags”
To Sum Up

- Pain is frequent
- Significant impact
- Contributions from vision, environment and personal factors
- Many can be mitigated
- Importance of self-efficacy
Where to from here?

• Extend this pilot study via online survey to get greater numbers

• Research into Vitamin D in PWA
  – Survey of knowledge and attitudes
  – Study looking at Vitamin D levels
www.surveymonkey.com/s/paininalbinism
References


• Bowling A. Just one question: If one question works, why ask several? J Epidemiol Community Health 2005;59:342-345 doi:10.1136/jech.2004.021204


Thankyou – Questions?