Managing complex cases of JDM into adult life

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Summary:

• What is normal adolescence?

• JDM in young adults

• Developmentally appropriate care for young adults with JDM
What is adolescence?
What’s a normal adolescence and young adulthood?

• Healthiest period in one’s life

• Life-changing events and ‘watersheds’

• Period of **immense change**
Psychological development in young adults?

....what is normal?
Normal development in adolescent brain:

- Pre-frontal cortex not fully developed until early 20s’
- ‘Executive suite’
  - Calibration risk/reward
  - Problem solving
  - Thinking ahead
  - Self-evaluation
  - Long-term planning
  - Regulation of emotion
Risk taking behaviour:

- Is it a good idea to swim with sharks?
- Is it a good idea to set your hair on fire?

Adapted from:
Reyna and Farley 2006
Baird and Fugelsang 2004
Risk taking in adolescence
How our capacity to lie peaks in young adulthood

The ability to lie follows a distinct pattern as we age, says Bruno Verschuere, who finds ways to spot liars in the hope of building better lie detectors.

Does our capacity to lie change?

There are age-related differences in our ability to lie, and these are in line with the development of the prefrontal cortex – a part of the brain involved in controlling our

(Image: Danny Schwarz)
Epileptic had stopped taking medication

AN EPILEPTIC was killed by a seizure after she refused to carry on taking her medication, an inquest has heard.
Katie Coombs, 20, had been diagnosed with the condition in 2009 but had stopped taking tablets and dodged out of seeing doctors. She died after suffering a fit which triggered a heart attack in August last year at her home in Didsbury, Greater Manchester.

The inquest heard she had missed several appointments to see a doctor last February and August.

Recording a verdict of death by natural causes, Stockport deputy coroner Joanne Kearsley said: ‘I don’t know why but since May 2010 she hadn’t been complying with her medication.’
Juvenile Dermatomyositis in young adults
Juvenile Dermatomyositis (JDM)

- Rare – 3.2 per million children per year
- Average age of onset – 7 years
- 25% ≤ 4 years
- F:M = 2.3:1

- Vasculopathy affecting:
  - Skin
  - Skeletal muscle
  - GI tract
  - Kidneys, eyes, heart
JDM – Diagnosis (Bohan and Peter 1975)

- Characteristic rash
  + 3 (for definite) or 2 (for probable) of the following:
  - Symmetrical proximal myopathy (80-100%)
  - Raised muscle enzymes (~90%)
  - Abnormal EMG (50-95%)
  - Abnormal muscle biopsy (75-90%)
JDM – heliotrope skin rash

BM Feldman et al, Lancet 2008
Gottron’s papules

BM Feldman et al, Lancet 2008

Nail fold capillary dilatation
<table>
<thead>
<tr>
<th></th>
<th>Juvenile Myositis</th>
<th>Adult Myositis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age/gender</td>
<td>7yrs – F&gt;M</td>
<td>30-50yrs – F&gt;M</td>
</tr>
<tr>
<td>Clinical features</td>
<td>calcinosis, lipodystrophy, cutaneous ulceration, <strong>no cancer signal</strong></td>
<td>ILD and myocardial involvement more common, <strong>30% cancer</strong></td>
</tr>
<tr>
<td>Pathogenesis</td>
<td>Humoral attack on muscle capillaries, upregulation of MHC Class I myofibres, pDC infiltration, <strong>type I IFN response</strong></td>
<td></td>
</tr>
<tr>
<td>Treatment</td>
<td>1) Steroids (PO+IV), MTX (s/c), IVIG , HCQ</td>
<td>2) AZA, MMF, Cyclophosphamide, anti-TNF, RTX</td>
</tr>
<tr>
<td>Antibodies</td>
<td>Juvenile Myositis associations</td>
<td>Adult Myositis associations</td>
</tr>
<tr>
<td>---------------------</td>
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<td>-------------------------------------</td>
</tr>
<tr>
<td>TIF1α</td>
<td>Skin disease (20-30%)</td>
<td>Cancer associated</td>
</tr>
<tr>
<td>MDA5</td>
<td>Skin (amyopathic)</td>
<td>Skin and lung (Japanese)</td>
</tr>
<tr>
<td>NXP2 (anti-MJ)</td>
<td>Calcinosis, muscle cramps</td>
<td>Lung and cancer</td>
</tr>
<tr>
<td>Anti-synthetase</td>
<td>5%</td>
<td>20-25%</td>
</tr>
</tbody>
</table>
LL – contacted by father of a girl with JDM

‘The problem is that she has turned 19 and so is transitioning. The help we have had from x Hospital (with support from X) has to now been good but she is falling between adult and children with nothing happening. I have written to x (paed hosp) and x (adult hospital) but have had no response and I am very concerned that we need to do something very soon.’
LL – 19 years old

• JDM since aged 2
• Severe skin involvement
• Managed initially with- Azathioprine, CyA, IVIg (oesophageal involvement)
• Moved to X hospital – changed to MTX
• Pred throughout
LL – 19 years old

• Worsening calcinosis since aged 10/11 yrs
• IV Pamidronate infusions
• Aged 14 life-threatening sepsis
• Off immunosuppression for 2 years – rapid progression of calcinosis
• Restarted Pred and MTX 20 mg/wk
LL – 19 years old

- Nailfold capillary dilatation
- Heliotrope rash
- Elbows and knees fixed flexion ≥90º
- Widespread calcification – trunk/prox limbs
- Unable to assess power
- Chest clear

- Growth delay – short stature (30 kg)
- Cachectic
LL – 19 years old

- Bloods
  - CK 120
  - Cr 17
  - ESR 51
  - Hb 10.9
  - ANA / ENA – negative
  - Echo – normal
  - HR-CT chest – no ILD
LL – 19 years old
LL – 19 years old
LL – 19 years old
LL – 19 years old
Management

• Reviewed by physio and OT

• Counseled
  – Pros and cons of anti-TNF
  – Infliximab via PICC or portacath
  – Pre-treat with antibiotics
### Treatment for calcinosis

<table>
<thead>
<tr>
<th>Calcium / phosphate modulators</th>
</tr>
</thead>
<tbody>
<tr>
<td>- Calcium channel blockers - diltiazem</td>
</tr>
<tr>
<td>- Bisphosphonates – pamidronate, alendronate</td>
</tr>
<tr>
<td>- Sodium thiosulfate (chelates free calcium)</td>
</tr>
<tr>
<td>- Aluminium hydroxide (decrease intestinal absorption $PO_4$)</td>
</tr>
<tr>
<td>- Probenecid ($PO_4$ excretion, decreases extracellular ATP)</td>
</tr>
</tbody>
</table>

TNF inhibitors – infliximab, adalimumab

- IVIG
- Abatacept
- Rituximab
- Colchicine
- Thalidomide
- Intra-lesional Depo-Medrone

Aggressive control of disease to prevent onset and progression is key

If suspect – X-ray to map and repeat 12 months later to monitor
## UCLH adolescent and young adult JDM Cohort (n=56)

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean age</td>
<td>22 yrs</td>
</tr>
<tr>
<td>Mean duration of disease</td>
<td>12.8 years</td>
</tr>
<tr>
<td>Ongoing skin disease</td>
<td>9 (16%)</td>
</tr>
<tr>
<td>Calcinosis</td>
<td>9 (16%) – 3 severe</td>
</tr>
<tr>
<td>Treatment free remission</td>
<td>30 (54%)</td>
</tr>
<tr>
<td>Treatment:</td>
<td></td>
</tr>
<tr>
<td>MTX</td>
<td>13 (23%)</td>
</tr>
<tr>
<td>HCQ</td>
<td>15 (27%)</td>
</tr>
<tr>
<td>MMF</td>
<td>5 (9%)</td>
</tr>
<tr>
<td>AZA</td>
<td>5 (9%)</td>
</tr>
<tr>
<td>IVIG</td>
<td>3 (5%)</td>
</tr>
<tr>
<td>Biologic</td>
<td>6 (11%)</td>
</tr>
</tbody>
</table>

[www.centre-for-adolescent-rheumatology.org](http://www.centre-for-adolescent-rheumatology.org)
Developmentally appropriate care for adolescents and young adults with JDM
Adolescents with chronic disease:

Bridging the gap Between Paed and Adult care
Healthcare change - challenge:

• Paediatric
  – Multidisciplinary
  – More time
  – Parent orientated
  – Active follow up
  – Time to extract info
  – Psychosocial support

• Adult
  – Physician orientated
  – Small team
  – No family support
  – No psychosocial support
  – Large volume
  – Less time
  – Patient orientated
Outcomes:

Interested in research?

Juvenile Dermatomyositis Cohort Biomarker Study and Repository (UK and Ireland) (JDCBS)

Research into childhood myositis.

Diseases that cause inflammation of the muscles (known as myositis) are rare but serious. The most common form of childhood myositis is juvenile dermatomyositis (JDM). JDM affects about 3 children in every million. Because myositis in children is so rare there is a lack of evidence for best ways to treat it. To overcome this, in the UK, a network of researchers, scientists, nurses, physiotherapists and doctors who work with children with myositis agreed to work together, and this has led to a large, powerful collection of cases of childhood myositis each with data and samples stored. This study is called the Juvenile Dermatomyositis Cohort Biomarker...
Paed Rheum assessment:

Childhood Myositis Assessment Score (CMAS)

Patient Reference Number: __________________________

1. Head Elevation (neck flexion): Item Score: ________
   0 = unable  4 = 60-119 seconds
   1 = 1-9 seconds  5 = >2 minutes
   2 = 10-29 seconds
   3 = 30-59 seconds  No. of seconds:...................

2. Leg raise/touch object: Item Score: ________
   0 = unable to lift leg off table
   1 = able to clear table but cannot touch object
   2 = able to lift leg high enough to touch object

3. Straight leg lift/duration: Item Score: ________
   0 = unable  4 = 60-119 seconds
   1 = 1-9 seconds  5 = >2 minutes
   2 = 10-29 seconds
   3 = 30-59 seconds  No. of seconds:...................

4. Supine to prone: Item Score: ________
   0 = unable. Has difficulty even turning onto side; able to pull arms under torso only slightly or not at all
   1 = turns onto side fairly easily; but cannot fully free arms and is not able to fully assume a prone position
   2 = Easily turns onto side; has some difficulty freeing arms, but fully frees them and fully assumes a prone position

Date of visit: __________________________

9. Floor sit: Item Score: ________
   Going from a standing position to a sitting position on the floor
   0 = unable. Afraid to even try. Even if allowed to use a chair for support. Child fears that he/she will collapse, fall into a sit or self-harm
   1 = much difficulty. Able, but needs to hold onto chair for support during descent (unable to unwilling to try if not able to use a chair for support)
   2 = some difficulty. Can go from stand to sit without using a chair for support but has at least some difficulty during descent. Descends somewhat slowly and/or apprehensively; may not have full control or balance as manoeuvres into a sit
   3 = No difficulty. Requires no compensatory manoeuvring

10. All-fours manoeuvre: Item Score: ________
   0 = unable to go from a prone to an all-fours position
   1 = barely able to assume and maintain an all-fours position
   2 = can maintain all-fours position with straight back and head raised (so as to look straight ahead). But cannot crawl forward
   3 = Can maintain all fours, look straight ahead and crawl forward
   4 = maintains balance while lifting and extending leg

11. Floor rise: Item Score: ________
   Going from a kneeling position on the floor to a standing position
Paed Rheum assessment:

* MMT: 0 = no muscle action, 1 = flicker of muscle action, 2 = muscle action with gravity counterbalance, 3 = muscle action against gravity, 4 = muscle action against gravity with some resistance, 5 = full muscle strength, 9 = not done

<table>
<thead>
<tr>
<th>Function of the Muscle</th>
<th>0–10 Scale</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>No Movement</strong></td>
<td></td>
</tr>
<tr>
<td><strong>MOVEMENT IN HORIZONTAL PLANE</strong></td>
<td></td>
</tr>
<tr>
<td>Moves through partial range of motion</td>
<td>1</td>
</tr>
<tr>
<td>Moves through complete range of motion</td>
<td>2</td>
</tr>
<tr>
<td>Moves to completion of range against resistance Or</td>
<td></td>
</tr>
<tr>
<td>Moves to completion of range and holds against pressure Or</td>
<td></td>
</tr>
<tr>
<td><strong>ANTIGRAVITY POSITION</strong></td>
<td>3</td>
</tr>
<tr>
<td>Moves through partial range of motion</td>
<td></td>
</tr>
<tr>
<td><strong>Test Movement</strong></td>
<td></td>
</tr>
<tr>
<td>Gradual release from test position</td>
<td>4</td>
</tr>
<tr>
<td>Holds test position (no added pressure)</td>
<td>5</td>
</tr>
<tr>
<td>Holds test position against slight pressure</td>
<td>6</td>
</tr>
<tr>
<td>Holds test position against slight to moderate pressure</td>
<td>7</td>
</tr>
<tr>
<td>Holds test position against moderate pressure</td>
<td>8</td>
</tr>
<tr>
<td>Holds test position against moderate to strong pressure</td>
<td>9</td>
</tr>
<tr>
<td>Holds test position against strong pressure</td>
<td>10</td>
</tr>
</tbody>
</table>
MMT8:

Neck Flexion:
Lying completely supine. Allow neck to flex about 45’
Add resistance to the forehead

Shoulder Abduction:
Abduct a straight arm to 90’. Add resistance to proximal to the elbow. Stabilise the body with the other hand.
MMT8:

**Neck Flexion:**
Lying completely supine. Allow neck to flex about 45’
Add resistance to the forehead

**Shoulder Abduction:**
Abduct a straight arm to 90’. Add resistance to proximal to the elbow. Stabilise the body with the other hand.
Elbow Flexion:
Support the arm at the elbow, keeping the arm close to the body. Flex the elbow to 45° and add resistance proximal to the wrist.

Wrist Extension:
Support the forearm, keeping the arm close to the body. Extend the wrist to 60°. Add resistance to the back of the hand.
<table>
<thead>
<tr>
<th><strong>Knee Extension:</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>In a sitting position, extend the knee fully, and then allow 5’ flexion (so the knee is not locked). Place the resistance proximal to the ankle. Keep the other hand on the knee to ensure the knee does not lock.</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th><strong>Hip Extension:</strong></th>
</tr>
</thead>
<tbody>
<tr>
<td>Lying prone, keep the pelvis flat. Flex the knee to 90’ and then lift the upper leg and knee off the bed 15’. Place the resistance proximal to the knee, use the other hand to stabilise the pelvis.</td>
</tr>
</tbody>
</table>
MMT8:

Hip Abduction:

Lying on their side, with slight extension at the pelvis and forward tilt at the trunk. Abduct the hip, keeping the knee straight. Stabilise the pelvis with one hand and add resistance proximal to the knee.

Ankle Dorsi-flexion:

In lying supine; dorsi- flex the ankle to 5’ keeping the knee straight. Add resistance to the dorsal aspect of the foot.
Don’t just talk about myositis

HEEADSSS 3.0
• Home
• Education
• Eating
• Activities
• Drugs and alcohol
• Sexual health
• Suicide/spirituality/sleep
• Social media/general safety

http://contemporarypediatrics.modernmedicine.com/contemporary-pediatrics/content/tags/adolescent-medicine/heeadsss-30-psychosocial-interview-

www.centre-for-adolescent-rheumatology.org
Communicating with young people

• Young person first and foremost
• Therapeutic alliance – engage them as central person
• Be curious, non-judgmental, open-ended questions
• Avoid clinics with more than 1 other person observing
• Don’t try to be cool!
Tips communicating with young people in clinic

• Young person first and foremost
• Therapeutic alliance – engage them as central person
• Be curious, non-judgmental, open-ended questions
• Avoid clinics with more than 1 other person observing
• Don’t try to be cool!
• Be frank, avoid authoritarian approach
• Introduce choice
• Examine patient fully at every consultation
• Not rushed - cannot do effectively in 10 minutes
• Introduce idea of seeing patient alone early and stage
• Continuity of care