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Managing complex cases of JDM into adult life

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University College London Hospitals NHS Foundation Trust

Great Ormond Street Hospital for Children NHS Foundation Trust

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Summary:

- What is normal adolescence?
- JDM in young adults
- Developmentally appropriate care for young adults with JDM

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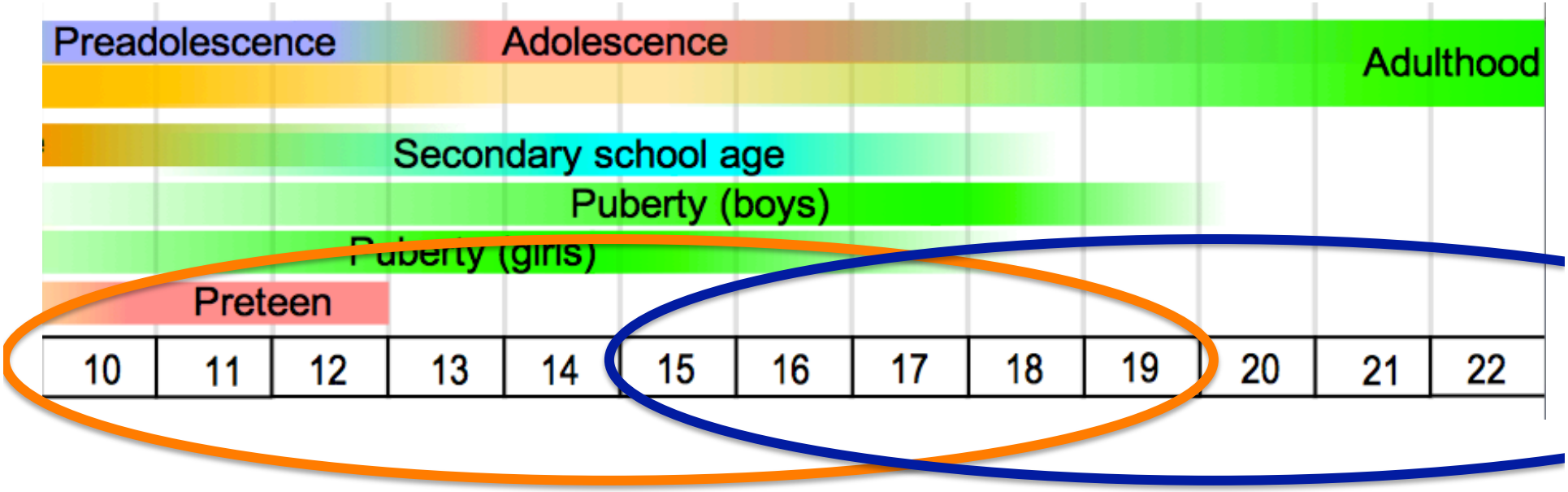
What is adolescence?


Preadolescence
Adolescence
Adulthood
Secondary school age
Puberty (boys)
Puberty (girls)
Preteen
10
11
12
13
14
15
16
17
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19
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21
22

What's a normal adolescence and young adulthood?

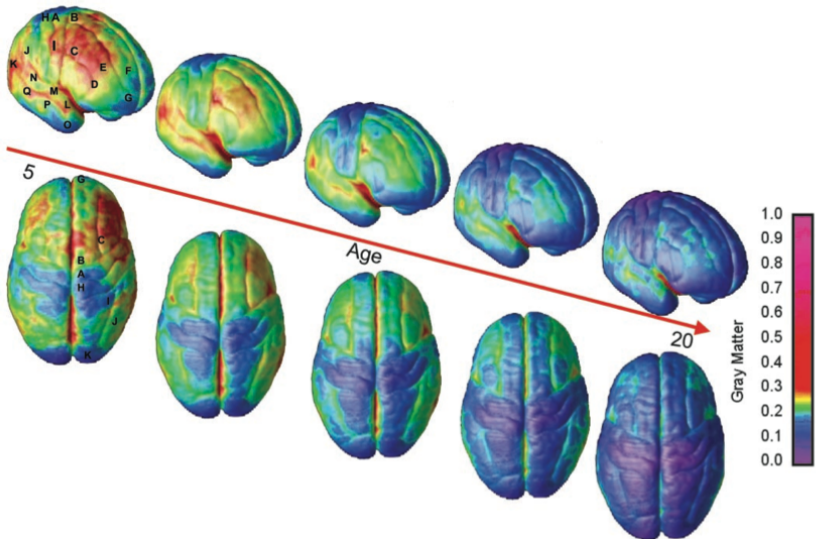
- Healthiest period in one's life
- Life-changing events and 'watersheds'
- Period of **immense change**

Psychological development in young adults?

….what is normal?

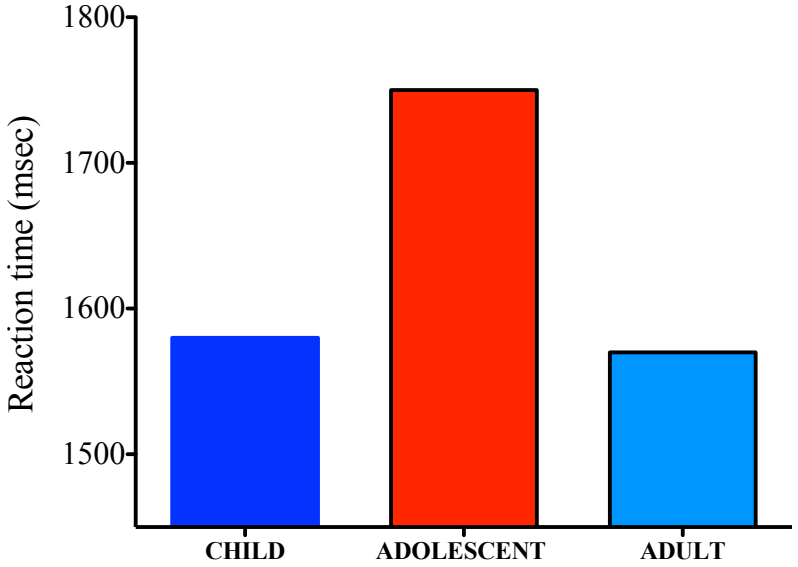
Normal development in adolescent brain:

- 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

Adherence

NewScientist
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OPINION 29 July 2015

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

(Image: Danny Schwarz)

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

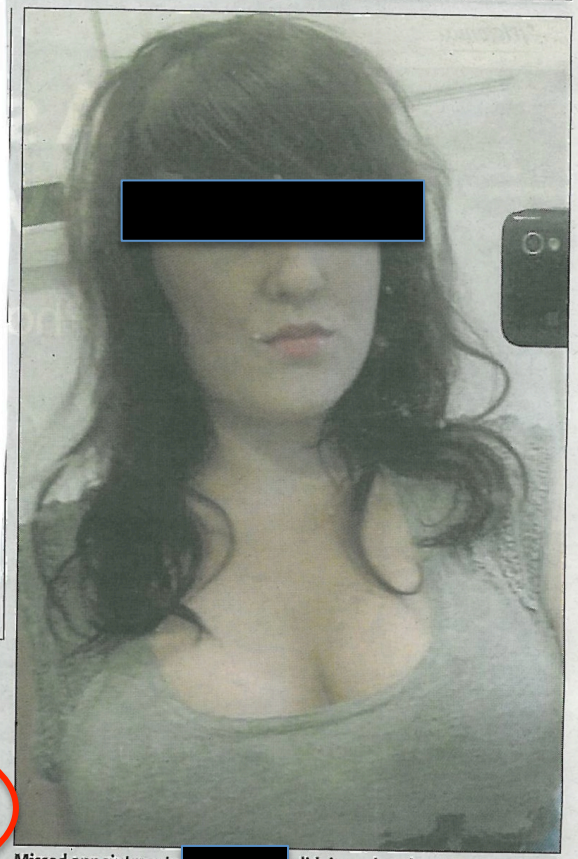
Epileptic had stopped taking medication

AN EPILEPTIC was killed by a seizure after she refused to carry on taking her medication, an inquest has heard.

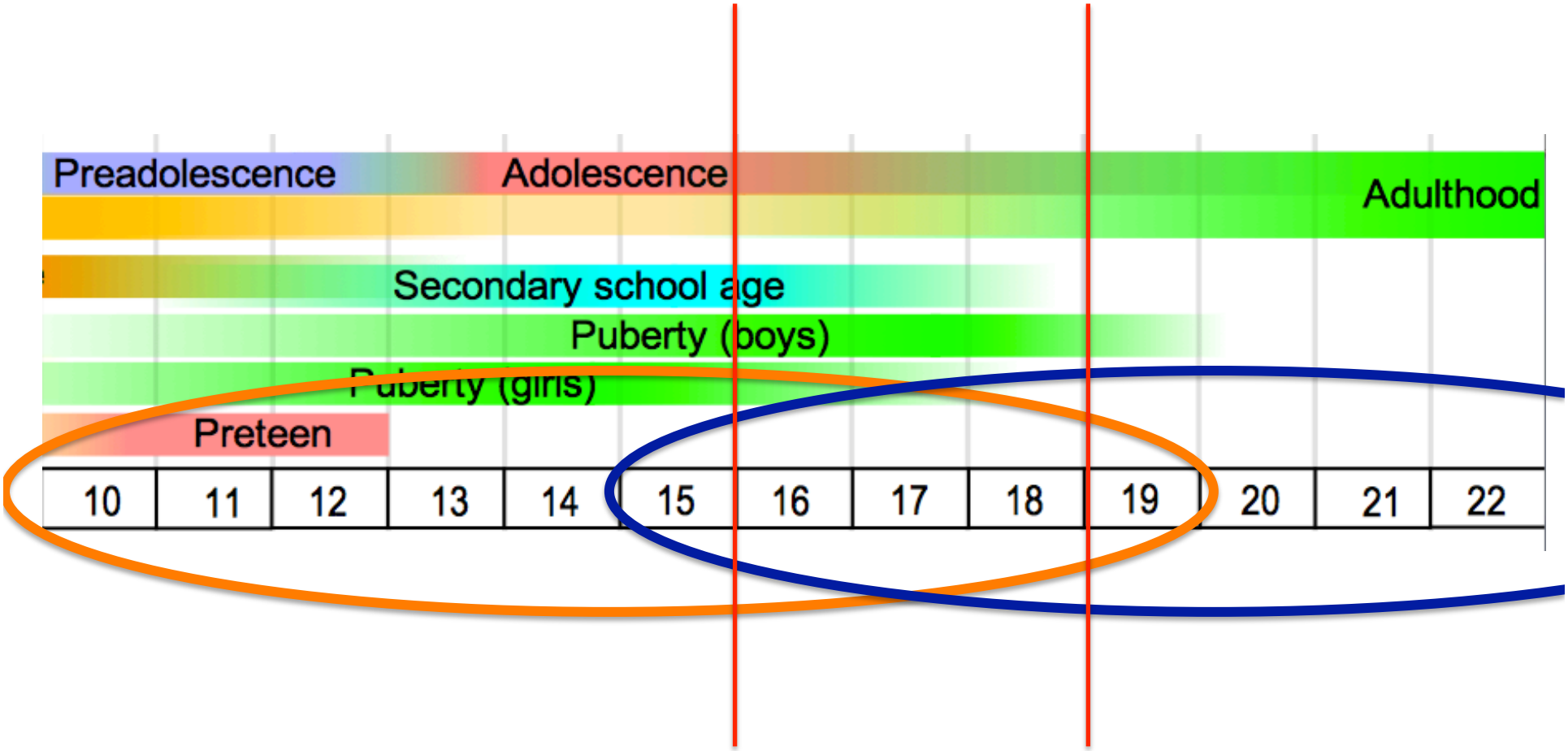
Katie Coombs, 20, had been diagnosed with the condition in 2009 but had stopped taking tablets and dodged out of seeing doctors. She died after suffering a fit which triggered a heart attack in August at her home in Droylsden, Greater Manchester.

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.'



Missed appointments: [redacted] didn't see her doctor PICTURE: CAVENDISH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Juvenile Dermatomyositis in young adults

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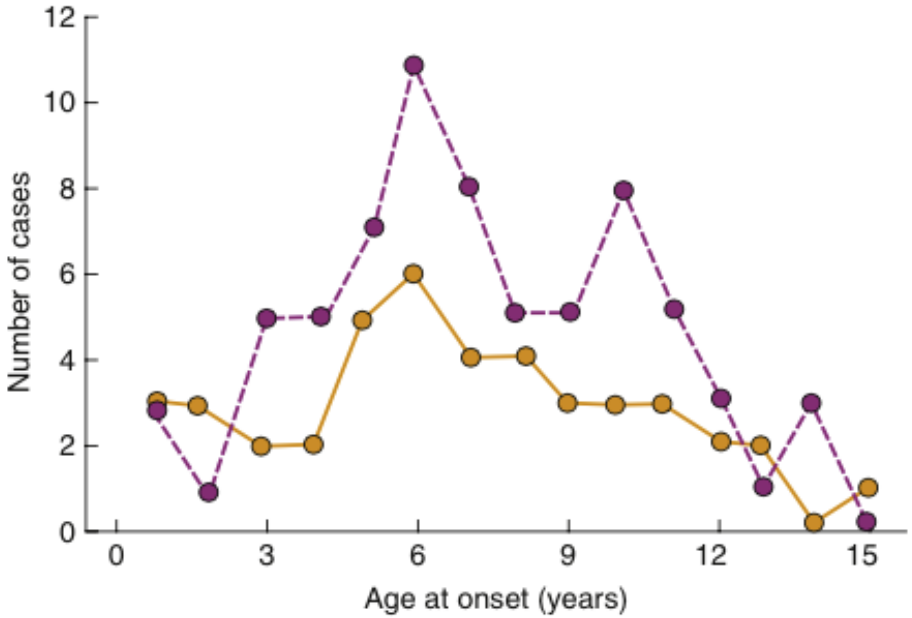
Juvenile Dermatomyositis (JDM)

- 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



Age at onset for girls *(dashed line)* and boys *(solid line)*.

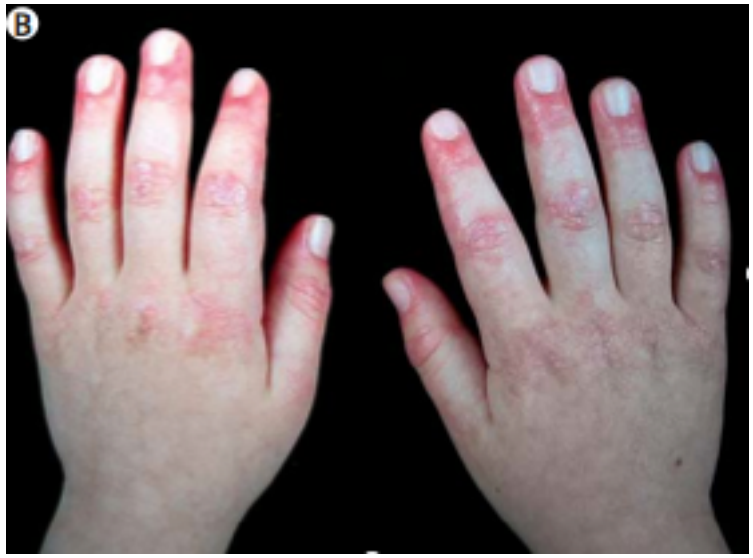
JDM – Diagnosis (Bohan and Peter 1975)

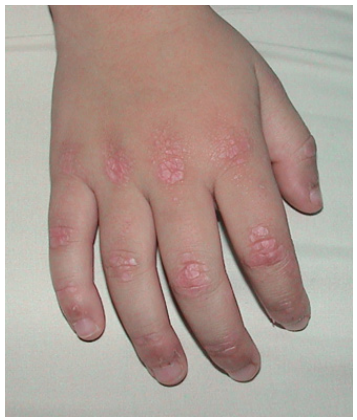
- **Characteristic rash**

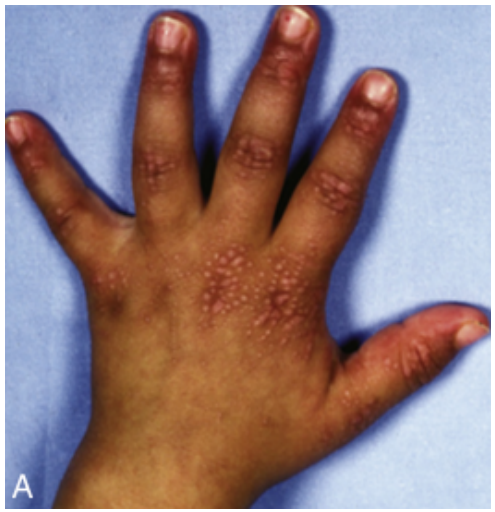
  + 3 (for definite) or 2 (for probable) of the following:

- Symmetrical proximal myopathy (80-100%)
- Raised muscle enzymes (~90%)
- Abnormal EMG (50-95%)
- Abnormal muscle biopsy (75-90%)

Gottron's papules

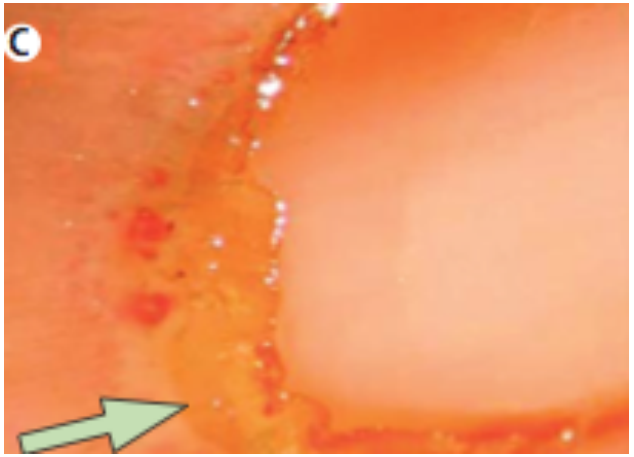


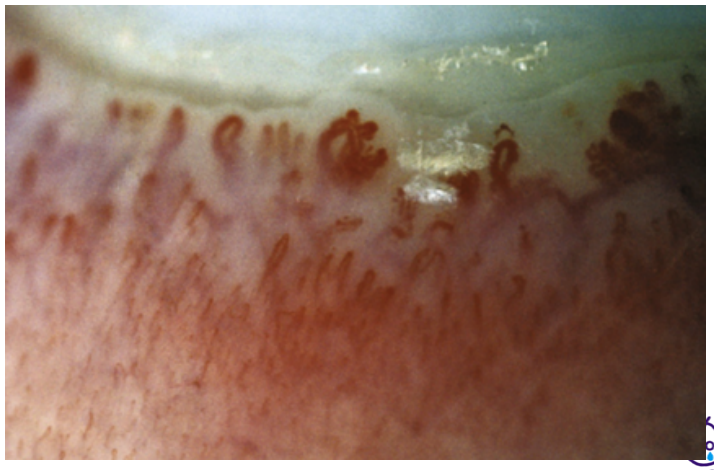




BM Feldman et al, Lancet 2008

Nail fold capillary dilatation





	Juvenile Myositis	Adult Myositis
Age/gender	7yrs – F>M	30-50yrs – F>M
Clinical features	**calcinosis**, lipodystrophy, cutaneous ulceration, **no cancer signal**	**ILD** and myocardial involvement more common, **30% cancer**
Pathogenesis	Humoral attack on muscle capillaries, upregulation of **MHC Class I myofibres**, pDC infiltration, **type I IFN response**	
Treatment	1) Steroids (PO+IV), MTX (s/c), IVIG , HCQ 2) AZA, MMF, Cyclophosphamide, anti-TNF, RTX	

Antibodies	Juvenile Myositis associations	Adult Myositis associations
TIF1α	Skin disease (20-30%)	Cancer associated
MDA5	Skin (amyopathic)	Skin and lung (Japanese)
NXP2 (anti-MJ)	Calcinosis, muscle cramps	Lung and cancer
Anti-synthetase	5%	20-25%

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

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

Calcium / phosphate modulators
- Calcium channel blockers - diltiazem
- Bisphosphonates – pamidronate, alendronate
- Sodium thiosulfate (chelates free calcium)
- Aluminium hydroxide (decrease intestinal absorption PO_4)
- Probenecid (PO_4 excretion, decreases extracellular ATP)

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)	
Mean age	22 yrs
Mean duration of disease	12.8 years
Ongoing skin disease	9 (16%)
Calcinosis	9 (16%) – 3 severe
Treatment free remission	30 (54%)
Treatment:	
MTX	13 (23%)
HCQ	15 (27%)
MMF	5 (9%)
AZA	5 (9%)
IVIG	3 (5%)
Biologic	6 (11%)

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Developmentally appropriate care for adolescents and young adults with JDM

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Adolescents with chronic disease:

Bridging the gap Between Paed and Adult care

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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

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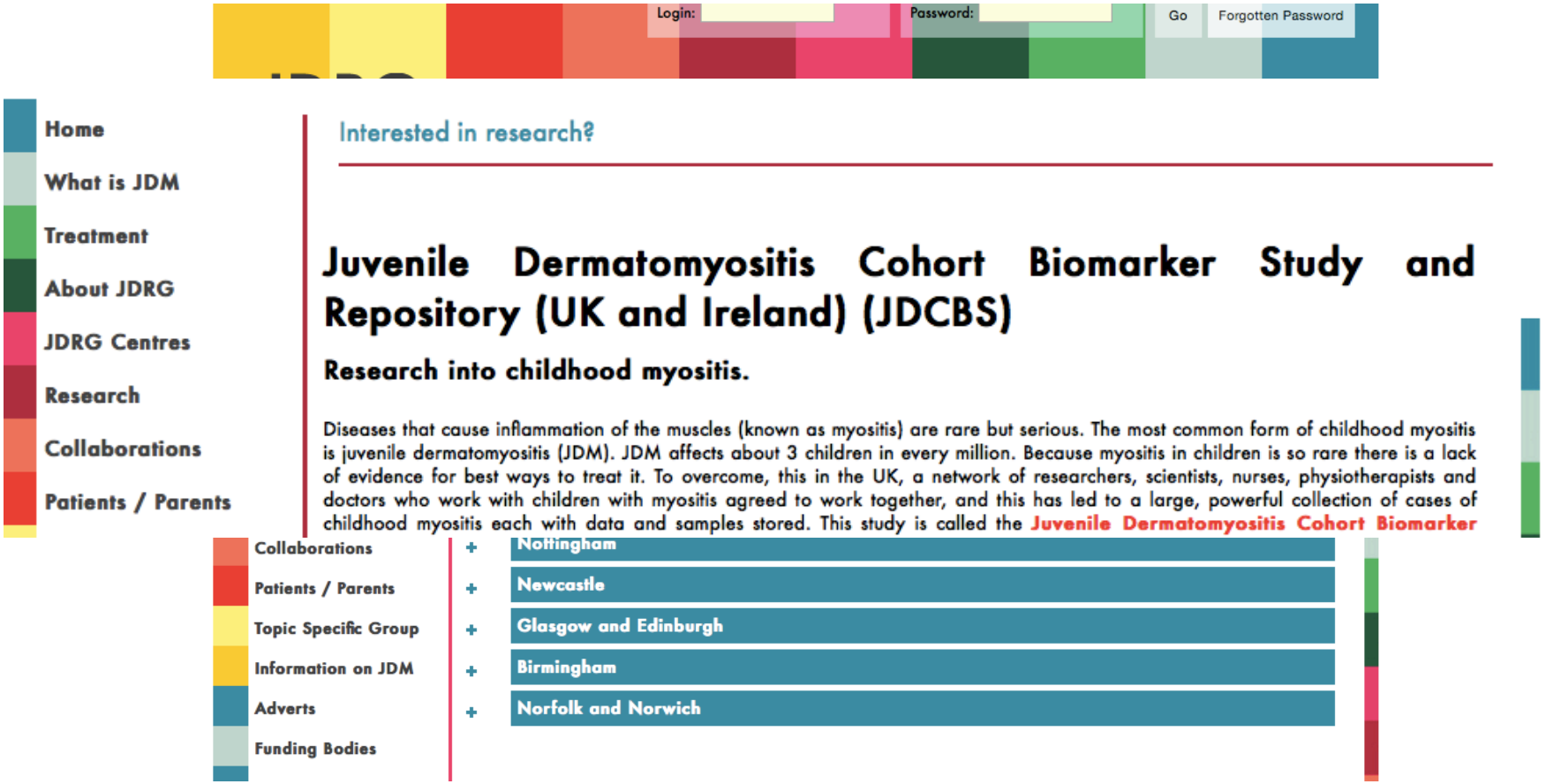
Outcomes:



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Paed Rheum assessment:

Childhood Myositis Assessment Score (CMAS)

Patient Reference Number:_____ **Date of visit:**_____

1. Head Elevation (neck flexion): **Item Score:**_____

0 = unable
1 = 1-9 seconds
2 = 10-29 seconds
3 = 30-59 seconds
4 = 60-119 seconds
5 = >2 minutes

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
1 = 1-9 seconds
2 = 10-29 seconds
3 = 30-59 seconds
4 = 60-119 seconds
5 = >2 minutes

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

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

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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)

STANDARD SCORE FOR Kendall MMT (0–10 SCALE)		
	FUNCTION OF THE MUSCLE	**0–10 SCALE**
No Movement		0
Test Movement	**MOVEMENT IN HORIZONTAL PLANE**	
	Moves through partial range of motion	1
	Moves through complete range of motion	2
	Moves to completion of range against resistance Or Moves to completion of range and holds against pressure Or	3
	ANTIGRAVITY POSITION	
	Moves through partial range of motion	
Test Position	Gradual release from test position	4
	Holds test position (no added pressure)	5
	Holds test position against slight pressure	6
	Holds test position against slight to moderate pressure	7
	Holds test position against moderate pressure	8
	Holds test position against moderate to strong pressure	9
	Holds test position against strong pressure	10

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Don't just talk about myositis

HEEADSSS 3.0

- **H**ome
- **E**ducation
- **E**ating
- **A**ctivities
- **D**rugs and alcohol
- **S**exual health
- **S**uicide/spirituality/sleep
- **S**ocial media/general safety

http://contemporarypediatrics.modernmedicine.com/contemporary-pediatrics/content/tags/adolescent-medicine/heeadsss-30-psychosocial-interview-adolesce?page=full

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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