Approach to Musculoskeletal Assessment

a.k.a a Rheumatologist’s search for the Holy Grail a story by Roger Allen
Genevieve

• 16 yr girl

• Elite gymnast (20+ hours/wk)

• FHx:
  – Mother – hypocomplementaemic vasculitis and hip arthroplasty

• Presents with L knee pain and swelling
  – Severe / Interferes with activity
  – No locking / instability
  – No trauma

• Referred to Ortho from Sports Med
• MRI (non contrast)
  – Diffuse large cystic mass anterior aspect of knee around infra patellar fat pad
  – diagnosis “ganglion”
  – also noted osteochondral defect lat tibial plateau

• Arthroscopy
  – significant chondral damage throughout knee
  – Marked synovitis
    • Histology c/w chronic synovitis
  – No bleeding noted
Rheumatology review:

- Further history:
  - Recurrent discomfort occurring over two year period
  - No morning stiffness
  - No limp
  - No locking

- O/E:
  - Warm knee
  - Boggy synovitis, no obvious effusion
  - Lack of full extension approx 10°
  - Marked quadriceps wasting
  - Otherwise well
Investigations

FBE, ESR, CRP - normal

HLA B27, RhF - negative

Ig’s, C3/C4 - normal

ANA 1:160
Therapy

• Intra-articular triamcinolone hexacetonide
  – “2 weeks” symptomatic improvement

• On review 4 weeks later:
  Increased pain ++
  Significantly reduced ROM ++
    Unable to flex beyond 40 deg
  No synovitis
  Tender tibial plateau lateral to patella tendon
• Repeat MRI (with contrast):
  
  – Severe arthropathy
    • Loss of both tibial and femoral articular cartilage
    • Cyst formation in tibial subchondral bone
    • Bony oedema
    • No enhancing synovitis seen with contrast
  
  – Venous malformation
    • Within Hoffa’s fat pad (at site of presumed “ganglion”)

• US
  
  – Vascular malformation with venous and arterial components
Sclerotherapy x3

- Some improvement but pain end of day
- Coaching gymnastics!
- Regained full ROM
- Persistent quad wasting
- ? Likelihood of arthroplasty as young adult
Abbey:

• Referred for assessment of “severe arthritis of fingers”
• no history of pain, swelling or EMS
• minimal interference of function
• general health otherwise normal
• short stature – approx 3rd %ile (as were parents)
Abbey:
Trichorhinophalangeal syndrome

- TRPS type 1  autosomal dominant
- chromosome 8q24.12
- phenotype - bulbous, “pear shaped” nose
  - thin, slowing hair
  - ulnar deviated proximal IP joints
- cone shaped phalanges on xray
Aspects of History:

1. Nature of onset ie acute, insidious, recurrent, symptom pattern

2. Timing of problem ie EMS, post-activity

3. Duration of problem ie days, months, years

4. Triggers eg recent infection, trauma, medication

5. Where does child/parent consider most symptomatic site(s) ie joint, enthesis, muscle, tendon sheath, bone
Aspects of History:

6. Thorough systems review ie head to toe

7. Degree of functional impairment – be conscious of possible inconsistencies

8. Family history ie autoimmune, genetic, pain, autoinflammatory

**Plus** the usual obligatory paediatric issues ie past history, development, vaccination etc
Aspects of Examination:

1. Examine ALL joints and not only site of presenting complaint

2. Aim to localise the maximal site of expected pathology ie joint capsule, adjacent bone or muscle, tendon or ligament insertions

3. Look for signs that reflect the history questions ie acute vs chronic

4. Be aware of normal age related “variants”

5. Examine for possible musculoskeletal aspects of systemic disease AND, if relevant, extraarticular features of primary arthropathies

6. Is there any apparent mismatch between history and examination ie are there features of a pain amplification issue
MSK examination should therefore include:

1. Joints – signs of inflammation (swelling, tenderness, range of movement)
2. Entheses
3. Tendon sheaths – including dactylitis
4. Gait – antalgic, limp, Trendelenberg, postural influences (plus neurology)
5. Muscles – tenderness, weakness
6. Spinal assessment – flexion (Schober test), scoliosis
7. Growth – general, pubertal, local issues

PLUS general review in view of importance of assessing for extra-articular features
Hands On
Practical advice on management of rheumatic disease

pGALS – A SCREENING EXAMINATION OF THE MUSCULO-SKELETAL SYSTEM IN SCHOOL-AGED CHILDREN

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# The pGALS musculoskeletal screen

**Screening questions**
- Do you (or does your child) have any pain or stiffness in your (their) joints, muscles or back?
- Do you (or does your child) have any difficulty getting yourself (him/herself) dressed without any help?
- Do you (or does your child) have any problem going up and down stairs?

<table>
<thead>
<tr>
<th>FIGURE</th>
<th>SCREENING MANOEUVRES (Note the manoeuvres in bold are additional to those in adult GALS)</th>
<th>WHAT IS BEING ASSESSED?</th>
</tr>
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</table>
| ![Image](image1.png) | Observe the child standing (from front, back and sides) | - Posture and habitus  
- Skin rashes – e.g. psoriasis  
- Deformity – e.g. leg length inequality, leg alignment (valgus, varus at the knee or ankle), scoliosis, joint swelling, muscle wasting, flat feet |
| ![Image](image2.png) | Observe the child walking and *'Walk on your heels' and 'Walk on your tiptoes'* | - Ankles, subtalar, midtarsal and small joints of feet and toes  
- Foot posture (note if presence of normal longitudinal arches of feet when on tiptoes) |
| ![Image](image3.png) | *'Hold your hands out straight in front of you'* | - Forward flexion of shoulders  
- Elbow extension  
- Wrist extension  
- Extension of small joints of fingers |
| ![Image](image4.png) | *'Turn your hands over and make a fist'* | - Wrist supination  
- Elbow supination  
- Flexion of small joints of fingers |
| ![Image](image5.png) | *'Pinch your index finger and thumb together'* | - Manual dexterity  
- Coordination of small joints of index finger and thumb and functional key grip |
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| ![Image](184x4) | 'Touch the tips of your fingers' | • Manual dexterity  
• Coordination of small joints of fingers and thumbs |
| ![Image](536x5) | Squeeze the metacarpophalangeal joints for tenderness | • Metacarpophalangeal joints |
| ![Image](536x5) | 'Put your hands together palm to palm' and 'Put your hands together back to back' | • Extension of small joints of fingers  
• Wrist extension  
• Elbow flexion |
| ![Image](536x5) | 'Reach up, “touch the sky”’ and ‘Look at the ceiling’ | • Elbow extension  
• Wrist extension  
• Shoulder abduction  
• Neck extension |
| ![Image](536x5) | 'Put your hands behind your neck' | • Shoulder abduction  
• External rotation of shoulders  
• Elbow flexion |
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<tr>
<td><img src="203x30.png" alt="Image" /></td>
<td>‘Try and touch your shoulder with your ear’</td>
<td>• Cervical spine lateral flexion</td>
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<tr>
<td><img src="517x510.png" alt="Image" /></td>
<td>‘Open wide and put three (child’s own) fingers in your mouth’</td>
<td>• Temporomandibular joints (and check for deviation of jaw movement)</td>
</tr>
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<td><img src="203x30.png" alt="Image" /></td>
<td>Feel for effusion at the knee (patella tap, or cross-fluctuation)</td>
<td>• Knee effusion (small effusion may be missed by patella tap alone)</td>
</tr>
</tbody>
</table>
| ![Image](203x30.png) | Active movement of knees (flexion and extension) and feel for crepitus | • Knee flexion  
• Knee extension |
| ![Image](203x30.png) | Passive movement of hip (knee flexed to 90°, and internal rotation of hip) | • Hip flexion and internal rotation |
| ![Image](203x30.png) | ‘Bend forwards and touch your toes?’ | • Forward flexion of thoraco-lumbar spine (and check for scoliosis) |
Practical Tip – when to perform pGALS in the Assessment

- Child with muscle, joint or bone pain
- Unwell child with pyrexia
- Child with limp
- Delay or regression of motor milestones
- The ‘clumsy’ child in the absence of neurological Disease
- Child with chronic disease and known association with MSK presentations
**RED FLAGS**
(Raise concern about infection, malignancy or nonaccidental injury)

- Fever, malaise, systemic upset (reduced appetite, weight loss, sweats)
- Bone or joint pain with fever
- Refractory or unremitting pain, persistent night-waking
- Incongruence between history and presentation (such as the pattern of the physical findings and a previous history of neglect)
Practical Tip – normal variants: indications for referral

- Persistent changes (beyond the expected age ranges)
- Progressive or asymmetrical changes
- Short stature or dysmorphic features
- Painful changes with functional limitation
- Regression or delayed motor milestones
- Abnormal joint examination elsewhere
- Suggestion of neurological disease or developmental delay

<table>
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<th>TABLE 1. Normal variants in gait patterns and leg alignment.</th>
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<tr>
<td><strong>Toe-walking</strong></td>
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<tr>
<td><strong>In-toeing</strong></td>
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<td><strong>Bow legs (genu varus)</strong></td>
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<tr>
<td><strong>Knock knees (genu valgus)</strong></td>
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<tr>
<td><strong>Flat feet</strong></td>
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<tr>
<td><strong>Crooked toes</strong></td>
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Beighton score = hypermobility assessment
Practical Tip – while performing the pGALS screening Examination

• Get the child to copy you doing the manoeuvres

• Look for verbal and non-verbal clues of discomfort (e.g. facial expression, withdrawal)

• Do the full screen as the extent of joint involvement may not be obvious from the history

• Look for asymmetry (e.g. muscle bulk, joint swelling, range of joint movement)

• Consider clinical patterns (e.g. non-benign hypermobility and Marfanoid habitus or skin elasticity) and association of leg-length discrepancy and scoliosis)
Practical Tip – when inflammatory joint disease is Suspected

• The lack of reported pain does not exclude arthritis

• There is a need to probe for symptoms such as
  – gelling (e.g. stiffness after long car rides)
  – altered function (e.g. play, handwriting skills, regression of motor milestones)
  – deterioration in behaviour (irritability, poor sleeping)

• There is a need to examine all joints as joint involvement is often ‘asymptomatic’
Investigations:

- FBE plus acute phase parameters - ESR, CRP
  - can have marked thrombocytosis
  - beware high ESR with few joints

- ANA (up to 20% +ve in “normal children”) non-predictive of specific CTD developing
  - RCH cut-off 1:160

- Rheumatoid factor (generally low yield)
  (anti-CCP abs: Cyclic Citrullinated Peptide - ? role)
Investigations (continued)

• immunoglobulins (approx 4% IgA deficient)
• HLA B27 (approx 10% Caucasians +ve)
• LFT’s - often elevated transaminases
• ferritin - disproportionate elevation in SoJIA ??MAS
• clotting / d-dimers - suggestive of MAS
• “diagnostic” joint aspirate – rarely useful
Imaging:

- Plain xray – erosions
  joint space width
  assess epiphyseal development

- Ultrasound – effusions (eg hip)
  tendon sheaths
  shoulders – rotator cuff impingement

- Bone scan/CT – usually for exclusion of other causes

- MRI – gadolinium contrast excellent for synovitis
  cartilage status and early erosions
Figure 2 - Dactyliitis, or "sausage digit," is seen in the toes of a child with psoriatic juvenile idiopathic arthritis.
Figure 3 Neonatal lupus
Still’s original patient (1897):
Juvenile Idiopathic Arthritis: Current Criteria

1. Systemic Onset

2. Oligo (Pauci) articular (1 to 4 joints)
   a) Persistent
   b) Extended (>5 after 6 months)

3. Seronegative Polyarticular (> 5 joints)

4. Seropositive Polyarticular (Rh Factor +ve)

5. Psoriatic

6. Enthesitis Related (HLA B27+ve)

7. Unclassified
Oligo(pauci)articular:
- approx 30%
- higher risk in young, female, ANA +ve
Systemic JIA
Polyarticular

- sero-ve >> +ve
- large/small jnt
- symmetrical distribution
skin/nail changes/dactylitis:

Psoriatic JIA:
Enthesitis-related

- male >> female
- usually B27 +ve
- sacroiliitis may follow but xray –ve
  - during paediatric/adolescent yrs
- may evolve into ank spond
Juvenile arthritis: Plenty of scope for optimism (hopefully)
- Orthognathic surgery: combined orthodontic / maxillofacial surgery
BUT....

Just when you thought everything was straightforward along comes a new direction of MSK disorders:

The world of the “autoinflammatory” diseases.
STING Associated Vasculopathy with onset in Infancy: SAVI = “interferonopathy”
Cryopyrin associated periodic syndrome: NOMID/CINCA = “inflammasomopathy”
BUT that’s enough for tonight – GO HOME!