



GPNI - PAEDIATRIC RHEUMATOLOGY

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PAEDIATRIC RHEUMATOLOGY
CLINICAL FELLOW



GOALS FOR TODAY

- Overview of our service
- Referral pathway
- 2 cases
- Q+A time with Dr Campbell and Dr McKenna, our Regional Paediatric Rheumatologists

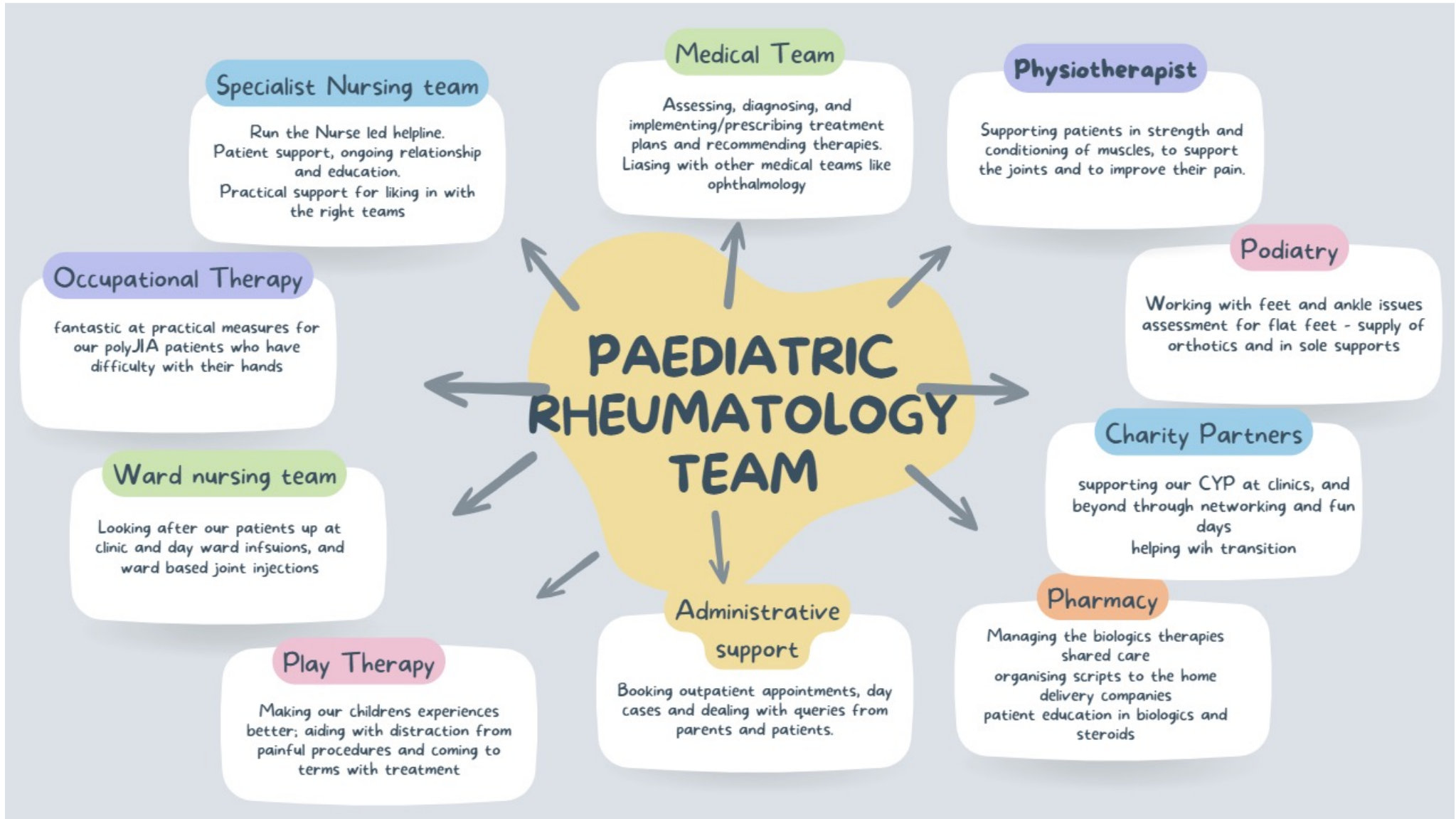


PAEDIATRIC RHEUMATOLOGY

- One tertiary service based in Musgrave Park Hospital, Belfast
- Usual route of referral from primary care is via general paediatrics
- Caseload of 1000+ patients at any one time
- >250 patients on biologic therapy
- >200 patients on DMARDS
- Predominantly outpatient based, in clinic and day ward
- Consult on inpatients also





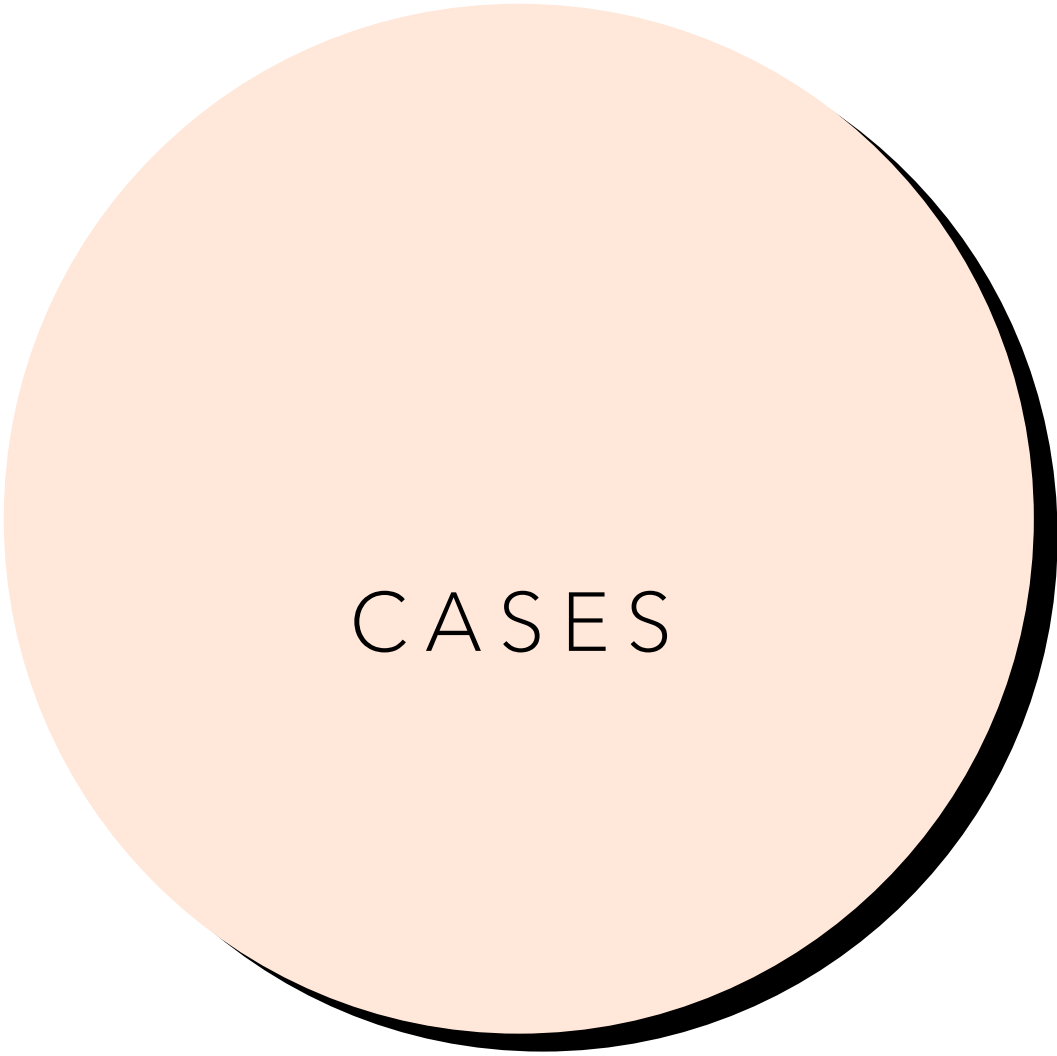




REFERRAL PATHWAY

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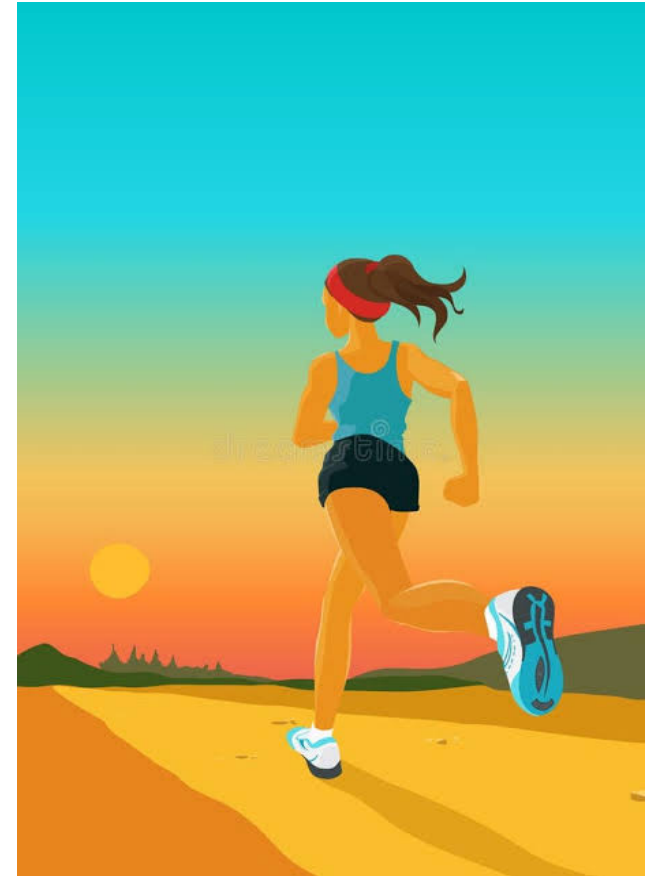
- Refer to general paediatrics
- Highlight that this could be a rheumatological issue
- They will see first and assess need for onward referral
- If you have concerns you can also contact us on the Registrar mobile - **07386670385**
- Concerns around new JIA should be made clear to allow timely assessment



CASES

PATIENT A

- 14 year old girl
- Usually fit and well
- Sporty - gaelic football and competitive swimming
- Presenting with back pain for the last 12 months
- Has tried physio but no great improvement
- Stiffness in the morning, and after sitting for a while
- Has had to give up her sports due to the pain



KEY POINTS IN THE HISTORY

- This pain persistent over time
- Stiffness in the morning and after periods of inactivity
- It's impactful – she has given up a very active lifestyle secondary to the pain
- No history of trauma, injury or prior illness.
- Family history – Mum has psoriasis and Maternal aunt has vasculitis, on biologic.
- Systemic enquiry – any rashes, fevers, night sweats, fatigue, weight loss?



CONTEXT

- Social – who lives with her?
- What activities does she like doing? Sport?
- What's the impact of the symptoms?
- What year of school – important exams?
- What subjects?
- Handwriting?

EXAMINATION FINDINGS

Look / Feel / Move approach

- Patient A has a slight loss of the usual lumbar lordosis
- She struggles with forward flexion of her spine
- She has almost no extension at the lumbar region
- Lateral spinal flexion reduced
- Pain with all these movements
- No rashes, muscle wasting, or scars.



EXAMINATION CONTINUED

- PGALS carried out - Paediatric Gait Arms Legs Spine
- 3 questions and then movements
- Any pain or stiffness in muscles, joints or back?
- Any difficulty getting dressed?
- Any difficulty going up and down stairs?
- **Examine Every Joint, Every time**



DIFFERENTIALS

- Muscular?
- Trauma?
- Oncology?
- Infection?
- Inflammatory arthritis?



INVESTIGATIONS IN PRIMARY CARE

- Working with our differentials, we want to make sure:
- Full blood count - cells lines appropriate, WCC,
- CRP - inflammation or infection
- U+E
- ESR for inflammation

INVESTIGATIONS

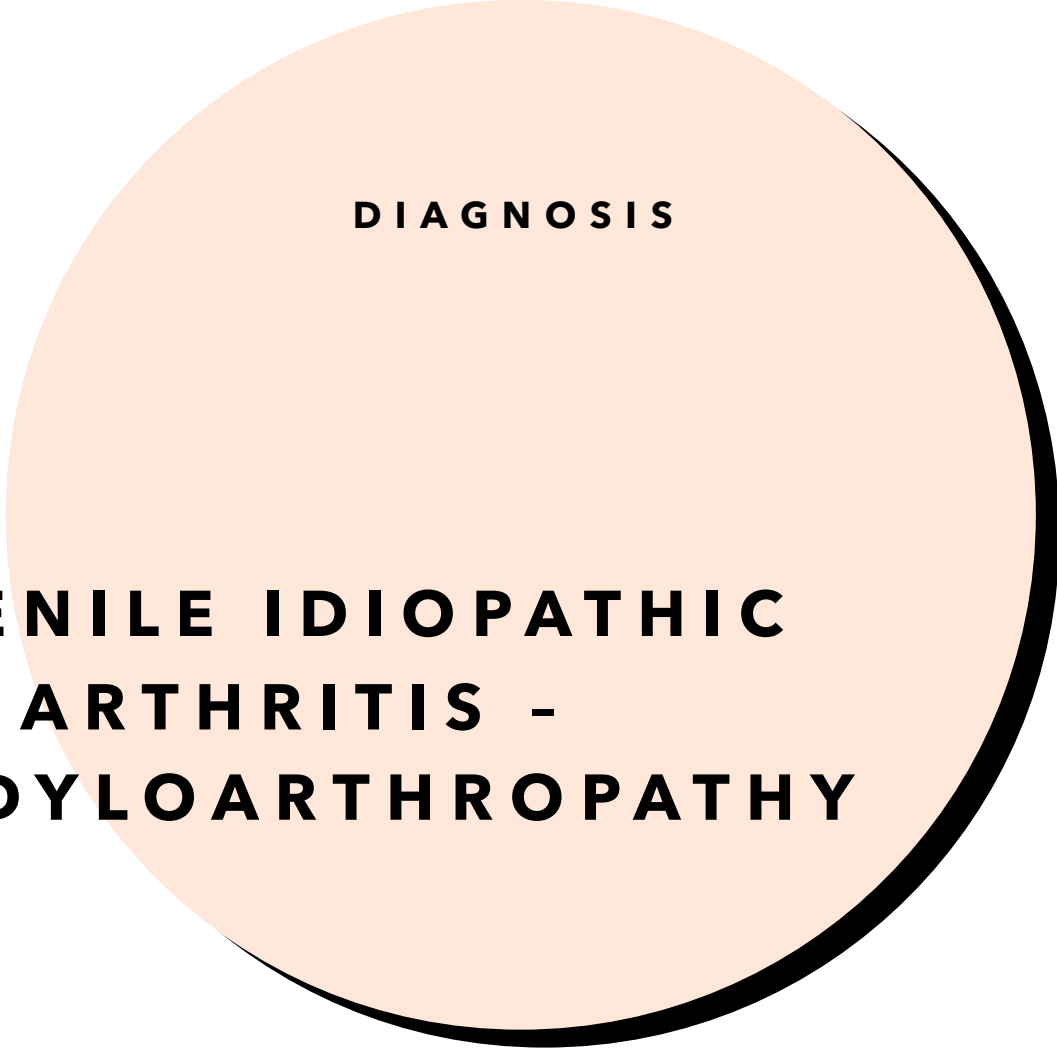
- Bloods - should be used to help confirm your diagnosis, not make it.
- Unlikely to be done in primary care
- CRP 0.7
- ESR 5
- FBC normal
- U+E/LFTs normal
- HLA-B27 was negative



INVESTIGATIONS

- MRI – Lumbar spine in the private sector orthopaedics
- Normal apart from a central schmorls node.
- Ongoing physio with no help, she went for further assessment
- MRI SI Joints showed bilateral effusions and enthesitis, with acute bilateral sacroileitis





DIAGNOSIS

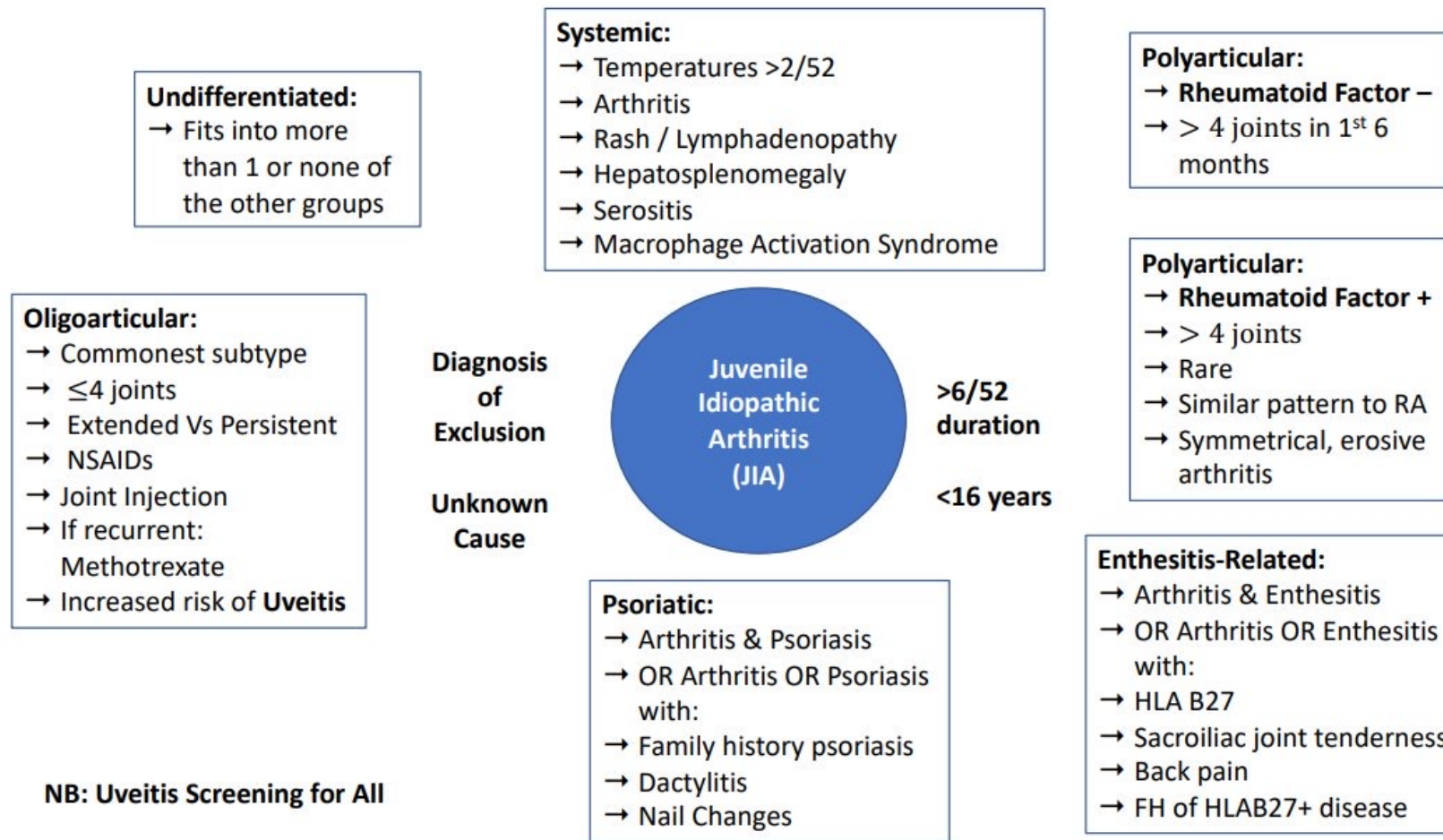
**JUVENILE IDIOPATHIC
ARTHRITIS -
SPONDYLOARTHROPATHY**

JUVENILE IDIOPATHIC ARTHRITIS

Umbrella term encompassing a complex range of inflammatory arthritis in patients;

- Under 16 years of age
- Present for at least 6 weeks
- Without another identifiable cause

Prevalence of around 4/1000 children



MANAGEMENT

- Patient A needs to go onto a biologic therapy.
- She has been symptomatic for >12months, and her life is very restricted.
- She starts on Adalimumab (Humira)



The goal of
management is
perfect joints

MANAGEMENT



- She undergoes pre-biologic screen to check for infection
- TB
- Hep C / Hep B
- HIV
- VZV
- Measles

MANAGEMENT

- She needs support and education about her disease from our specialist nurses.
- She gets education about Humira from our Pharmacist.
- She gets assessed and given stretches and movements to do from physio.
- She links in with the team from Arthritis UK who help her meet other young people with JIA
- She is referred to ophthalmology for Uveitis Screening

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2 MONTHS LATER

- Using Naproxen for pain relief and her back is starting to feel a little better
- Patient A has a temperature and a sore throat. She is fatigued and feels rotten.
- She phones our nurse advice line
- Our specialist nurses advise her that she needs to be assessed to decide if she should continue to take her Humira, which is due in 2 days time.
- She goes to see her GP.



How It Works:

When you call the Advice Line, you will speak with a member of our admin team. They will ask you for some information including:

- Your child's full name and date of birth
- Your child's health and care number (if you have it)
- The reason for your call.

Telephone Advice Line
07526202993

General Admin queries
(appointments, letters, results)
02895043973

Please do not send any queries via email unless a member of staff has asked you to do so.

Useful Links:

Arthritis UK

www.arthritis-uk.org

National Rheumatoid Arthritis Society (NRAS)

www.jia.org.uk

CCAA - Kids with Arthritis

www.ccaa.org.uk

Lupus UK

<https://lupusuk.org.uk>

Myositis UK

www.myositis.org.uk

Behcets UK

<https://behcetsuk.org>

Vasculitis UK

www.vasculitis.org.uk

RACC (Autoinflammatory Diseases)

<https://raccuk.com>

Belfast Health and Social Care Trust

<https://belfasttrust.hscni.net>



Belfast Health and
Social Care Trust

caring supporting improving together

Nurse-led Telephone Advice Line



**Paediatric
Rheumatology**
— Northern Ireland —

**Information for
Patients**

Paediatric Rheumatology Nurse-led Telephone Advice Line

We offer a nurse-led telephone advice service for Paediatric Rheumatology patients and their relatives only.

**Monday - Friday
9:30am - 12:00pm**

**Telephone Advice Line
07526202993**

The service is not available on Public Holidays.

The advice line is **NOT** an emergency line.

Your call will be returned within **72 hours** and if not please ring again.



If you need urgent advice, or advice not related to your rheumatology condition, you should contact your GP, attend your nearest Emergency Department or in an emergency dial 999.

When to call:

- ✓ If your child has a 'flare up' of their condition and you need advice.
- ✓ If your child is experiencing side effects which you feel may be caused by their rheumatological medication.
- ✓ If you have a query regarding vaccinations.

When NOT to Call:

- ✗ To change or cancel an appointment or request letters, please contact General Admin queries.
- ✗ To obtain test results, please contact General Admin queries.

GP ASSESSMENT

- Fever 38.3
- Painful swallow
- Swollen lymph nodes
- Rapid Strep test is positive
- What do we need to do?



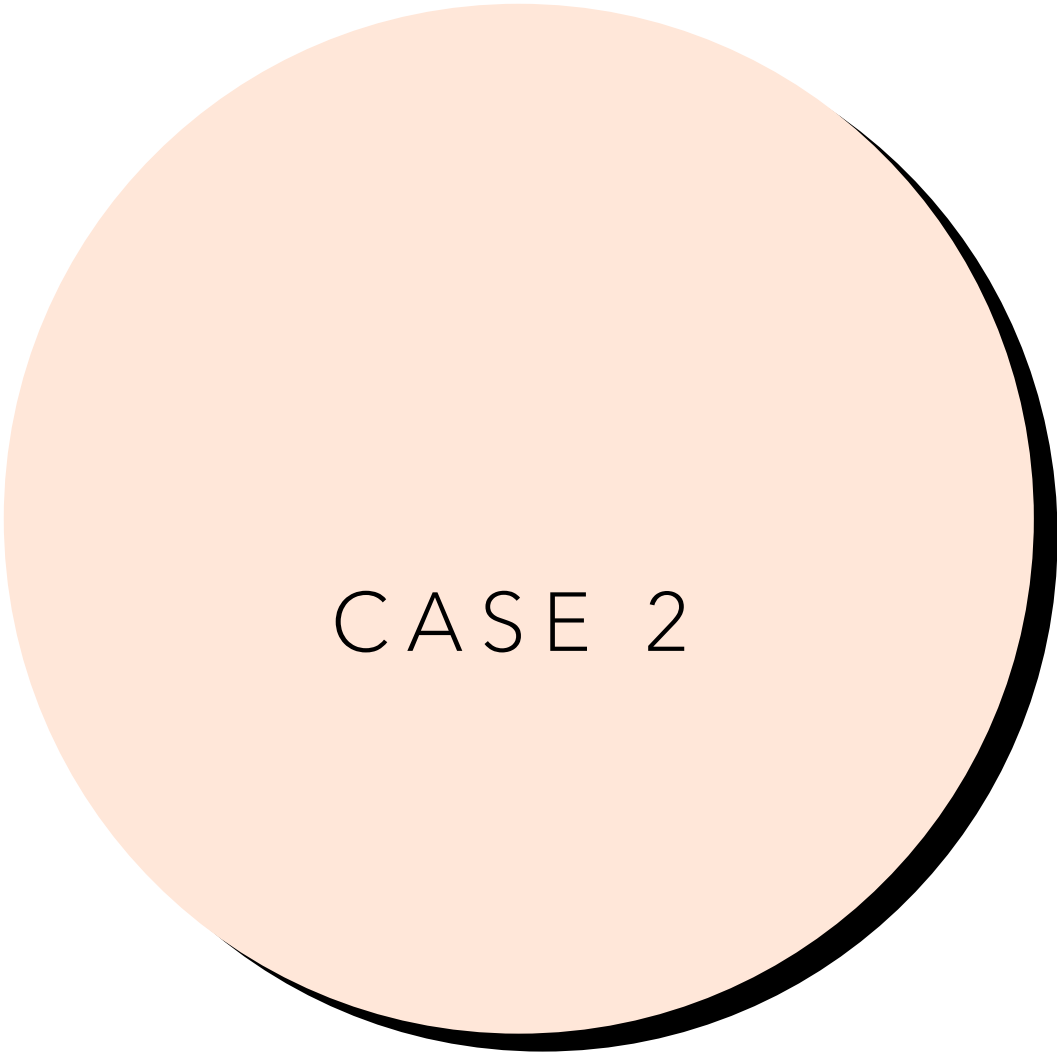
CONSIDER THE ADALIMUMAB

If you have concerns over a bacterial infection, then we need antibiotics.

Adalimumab should be held until

- fever has settled **and**
- antibiotic course is complete
- Be mindful that what is a common illness to you or me, is potentially more dangerous in kids on immunosuppression



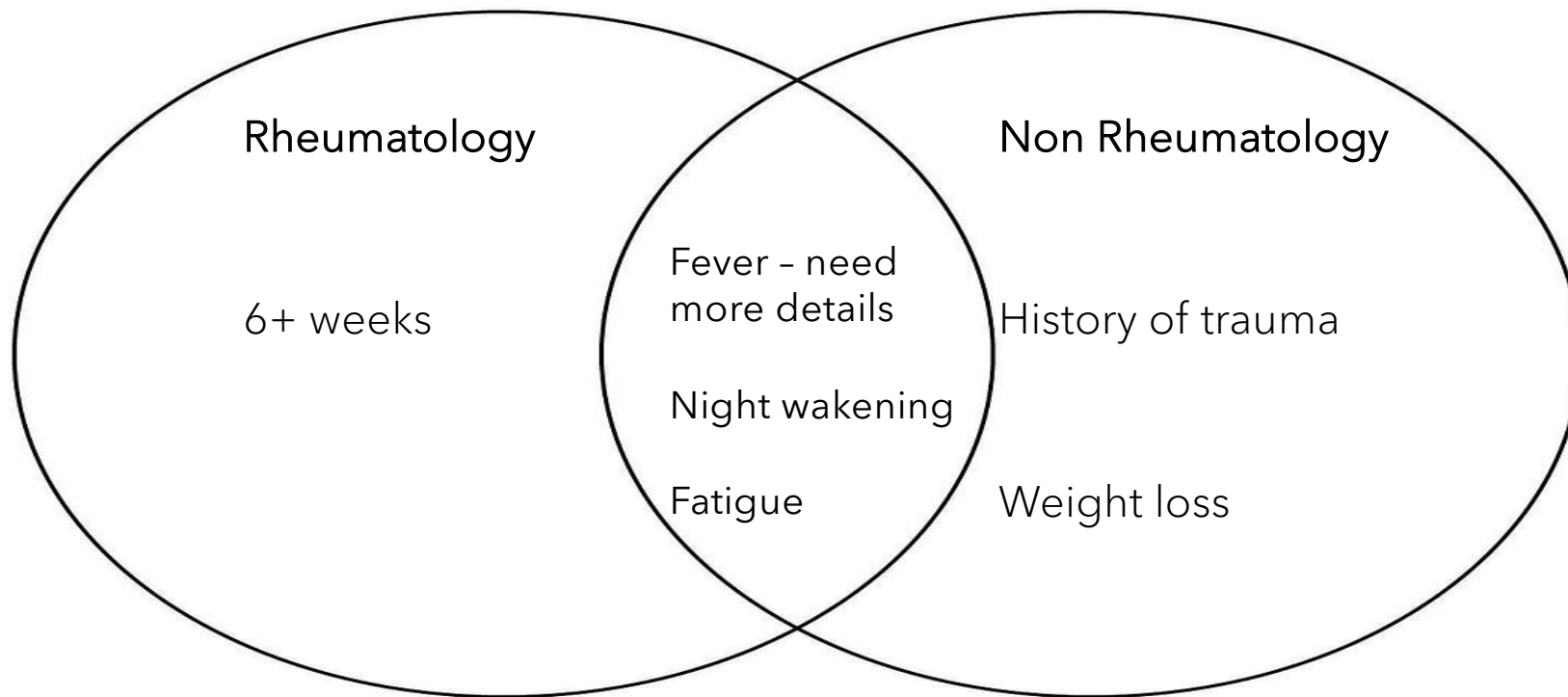


CASE 2

PATIENT M

- 6 year old girl, usually fit and well.
- History of minor trauma to right ankle – went over on it and sprained it July
- Has been sore on and off since
- Further R ankle injury on a bouncy castle mid-August
- Had 4-5 febrile illnesses during the next 6 weeks
- Attends the GP in September with pyrexia - 2 days fever, some weight loss, fatigue, **and** progressively non weight-bearing and complaining of pain in her right ankle.
- Wakening overnight complaining of pain in foot and tummy

WHAT ARE THE KEY FEATURES IN THE HISTORY?



DIFFERENTIAL DIAGNOSES

- Infection/ sepsis
- Septic joint
- New oncology diagnosis
- Reactive arthritis (4-6 weeks)
- NAI
- Skeletal dysplasia



WHAT NEXT?

WHAT ABOUT BLOODS?

- If you saw this patient in primary care, would you do bloods?
- If so, what would you send?



BLOODS THAT ARE USEFUL

- Full blood count
 - CRP
 - U+E
 - ESR
-
- If worried about a septic joint - don't waste time doing bloods- just send to ED



SENT TO ED - EXAMINATION

- Pale, but alert and chatty
- Chest is clear, no creps or increased work of breathing
- Cardiac; has a soft murmur, pulses good, CRT <2seconds
- Abdomen soft, non tender, no masses
- ENT; Ears NAD, Tonsils large, no exudate
- No rashes, no meningism
- Walking with intermittent limp
- Right ankle; some discomfort on movement, tender over 3rd metatarsal, Neurovascularly intact. No erythema or swelling.



WHAT NEXT?

Examine every joint, Every time.



WORKUP

Routine bloods to start

- CRP 230
- ESR 126
- HB 99
- WCC 10
- PLT 722

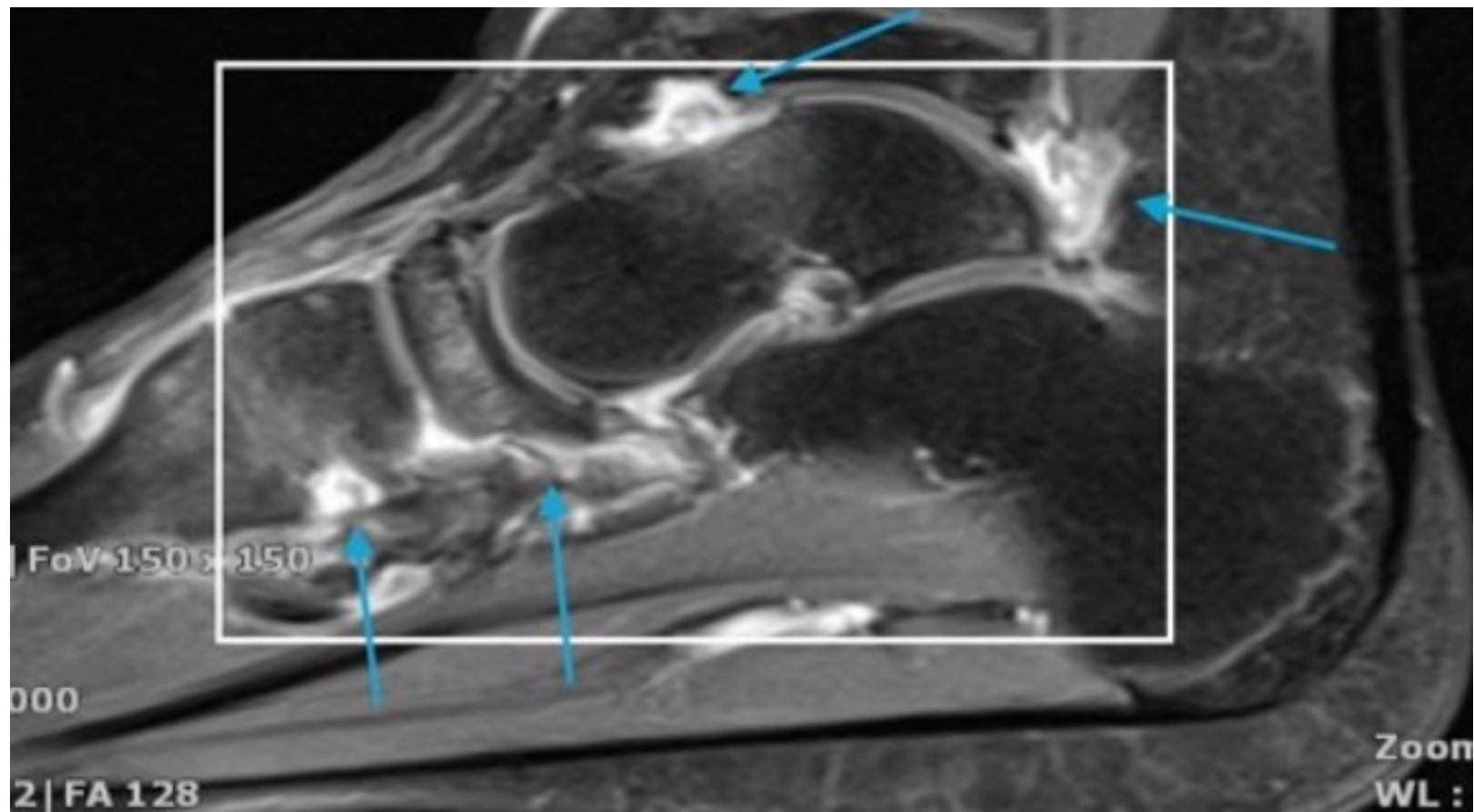


ADMISSION FOR IV ANTIBIOTICS



- Reviewed with orthopaedics for ?septic joint
- Ortho considering surgical intervention
- Progressively non-weightbearing
- MRI foot and ankle

MRI



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RHEUMATOLOGY INVOLVEMENT

- Examination of all her joints revealed that her knee extension wasn't great bilaterally
- Left ankle also felt a little hot
- Bedside ultrasound –
- Right ankle effusions at Main joint, subtalar and tendon involvement
- Effusions bilaterally in the knees
- Activity around the left main ankle joint

Diagnosis: **POLYARTICULAR JIA**



PATIENT M'S MANAGEMENT

- She had her baseline bloods completed
- Her disease activity was florid so she got pulsed with high dose IV Methylprednisolone for 3 days, then weaned to oral steroids.
- Like our first patient, she was educated by our specialist nurses
- She got input from our play therapist (needle-phobic)
- Needed assessment and advice from physio, and podiatry due to foot being affected

STEROID WEANING PLAN

Oral steroids for home - see steroid weaning plan below:

Steroid Dose	Length of time
30mg	7 days
25mg	7 days
20mg	7 days
15mg	7 days
10mg	7 days
5mg	7 days

		Tick (✓)
1.	Patient/Carer demonstrates understanding of the reason why Prednisolone has been prescribed.	Yes
2.	Understanding of how Prednisolone is taken.	Yes
3.	Understand if Prednisolone is used for more than 7 days the dose will gradually be reduced over a certain period of time.	Yes
4.	Understand why steroids should never be stopped suddenly .	Yes
5.	Understand if steroid therapy is greater than 4 weeks, patient may require calcium and vitamin D supplements.	Yes
6.	Understand what patient should do if there is a missed or vomited dose of Prednisolone.	Yes
7.	Aware of common side effects of Prednisolone: • May cause irritation to the stomach lining. • Sleep can be disrupted. • Increased appetite. • Increased risk of infection. • High blood pressure. • Changes in mood or behaviour.	Yes
8.	Patient should be provided with the following information on commencement of prednisolone treatment: • Steroid treatment card (blue). • Leaflet 'Your Child's Prednisolone' (includes contact details for paediatric rheumatology team). • Prescription for Prednisolone will be sent electronically to GP surgery.	Yes

METHOTREXATE

- Patients and parents will be trained in administration
- Usually aim for Friday
- Main side effects NAUSEA, GI upset, mouth ulcers, LFT derangement, cytopenias
- Folic acid helps with this
- Shared care guidelines



Monitoring table		Hospital specialist	GP		Hospital specialist
Test	Indication	Pre-treatment baseline	During treatment		At review
			Until on stable dose for 6 weeks	Thereafter	
FBC	Baseline assessment, dose adjustment	✓	Every 2 – 4 weeks (as advised by specialist)	Every 3 months	As part of review or as clinically indicated
LFTs					
U&Es, eGFR					
ESR/CRP (Rheumatology and Gastroenterology only)	Disease activity scoring	✓	Every 3 months		If clinically indicated
Height & weight	Baseline assessment	✓	Not routinely required		
PIIINP (Dermatology only)					
Blood pressure	Baseline assessment, respiratory and TB screening	If clinically indicated			
Chest x-ray					
PFTs, TB screening if indicated					
Ask about oral ulceration, sore throat, unexplained rash or unusual bruising/bleeding		✓	At every consultation		✓
If a further DMARD /JAK is added as combination therapy, or the dose is increased, the initial starting schedule should be reinstated. There may be clinical circumstances where the frequency of monitoring may vary and this should be specified by the initiating specialist.					

GP responsibilities

- Prescribe methotrexate (dose and device/brand as specified by hospital specialist) once each week (specify day not Monday). “As required” or “as directed” are **unsuitable** dosage instructions for subcutaneous methotrexate. [See MHRA advice on preventing inadvertent daily dosing.](#)
- Prescribe folic acid as specified by hospital specialist.
- Prescribe Sharpsguard or Sharpsafe container 1 litre (each will hold up to four pens).
- Arrange and record ongoing monitoring as advised by specialist (see monitoring table), ensuring practice systems are in place to recall patients for monitoring blood tests.
- Follow-up any non-compliance with the monitoring schedule. The risks of cessation of therapy versus risks of toxicity should be considered. Contact the specialist if treatment is stopped or further advice required.
- Report any adverse drug reactions to initiating specialist and the usual bodies (eg. MHRA/CHM).
- Ensure no drug interactions with other medicines.
- Check patient is using adequate contraception if appropriate.
- Administer **inactivated** influenza vaccine annually unless otherwise advised by the initiating specialist. Note the live formulation (eg. Fluenz Tetra[®]) must not be used.
- Check patient has received pneumococcal vaccine according to BNF or Green Book schedule.
- Provide COVID 19 vaccination as appropriate as per local arrangements and Green Book.
- Post exposure prophylaxis (antivirals or VZIG if antivirals are contraindicated) should be considered in non-immune at risk patients if exposed to chickenpox or shingles. Contact the consultant virologists, Regional Virus Laboratory, Royal Group of Hospitals on 07889 086 946 for advice if exposure is suspected. For other queries eg. those concerning exposure, infection or any recommendations relating to healthy susceptible household contacts, consult the Green Book and/or take additional advice from Regional Virus Laboratory, Royal Group of Hospitals
- Ask about oral ulceration, sore throat, unexplained rash or unusual bruising/bleeding at every consultation.

Withhold methotrexate and contact specialist if:

- WCC < $3.5 \times 10^9/L$
- Neutrophils < $1.6 \times 10^9/L$
- **Unexplained** eosinophilia > $0.5 \times 10^9/L$
- Platelets < $120 \times 10^9/L$
- MCV > 105fL, (check B12 & folate & TFT)
- AST/ALT > 3 times the upper limit of normal (for results between 2 - 3 x ULN, continue methotrexate, repeat bloods and seek specialist advice) Minor elevations of AST/ALT are common
- If renal impairment develops (not always appropriate to stop but may need dose adjustment)
- Unexplained fall in serum albumin
- Oral ulceration / sore throat
- Unexplained rash / abnormal bruising
- New or increasing dyspnoea or dry cough
- Persistent diarrhoea (can be a sign of toxicity).

Normal reference range may vary slightly between labs.

Please note an unusual fall or rise or a consistent downward or upward trend in any value should prompt review of the patient and extra vigilance. Some patients may have abnormal baseline values, specialist will advise.

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USEFUL WEBSITES

- Arthritis-UK.org (formerly versus arthritis)
- JIA.org.uk
- Pmmonline.org

