Commissioner's Guide to the NCEPOD Report 'Joint Care?'

A review of the quality of care provided to children and young adults with juvenile idiopathic arthritis (JIA)

Introduction

Juvenile idiopathic arthritis (JIA) is an autoimmune disease that affects around 10,000 children under 16 years of age in the United Kingdom.[1] It is a chronic disease and although it is diagnosed before a patient's 16th birthday, many patients will continue to have JIA into adulthood. JIA causes inflammation, pain and stiffness in joints, and can be debilitating. JIA is an umbrella term for a diverse group of conditions characterised by chronic arthritis and categorised into subtypes. The most common of which affects a small number of joints, but other types exist which can affect multiple joints, which is more likely to extend into adulthood, or be associated with other systemic diseases.

The pathway for children and young people diagnosed with JIA varies depending on the initial presenting symptoms as well as the organisational and commissioning arrangements of the treating hospitals including clinical networks and geographical location. Differential diagnosis for a painful joint is wide and although the red flag symptoms for an inflammatory arthritis are clear, patients are frequently referred to other specialties prior to rheumatology, such as paediatric orthopaedics or emergency medicine, or parents seek advice from physiotherapy services. This leads to unnecessary delays in diagnosis and treatment resulting in pain and an increased risk of joint damage.

Medical treatment for JIA is focused on suppression of the inflammatory response. Single joints can be treated with intra-articular (IA) steroid injections. IA injections frequently require administration under general anaesthetic or conscious sedation, in an appropriate setting with access to radiological support if required. They can also be given with topical anaesthetic and Entonox if tolerated. Access to slots with appropriate anaesthetic support is essential if therapy is to be given in a timely fashion.

When multiple joints are affected, intravenous steroids and/or oral corticosteroids are used initially before progressing to methotrexate. There is an existing medication pathway for JIA,[2] but this pathway does not include all currently available treatments and varies considerably around the UK leading to inequalities of access to appropriate therapy. If the disease is resistant to these treatments, then biologic medications are becoming available, and the National Institute for Health and Care Excellence has published technology appraisal guidance on their use.[3]

Patients receive most of their treatment at home, it is therefore important for them and their parents/carers to understand the risks and benefits of the medications, particularly methotrexate and to be confident in administering it. Methotrexate frequently has side effects making it difficult to tolerate. It is most often given by the subcutaneous route which requires training for patients and parent carers, the organisation of which can result in delays to treatment starting. The provision of training is variable and can lead to delays in starting therapy or increased risk.

Medications can also cause immunosuppression so there needs to be awareness of the signs and symptoms of sepsis and infections such as chickenpox. Additionally, methotrexate causes birth defects, so it is essential that sexual activity and birth control are discussed with the patient prior to treatment. These conversations need careful consideration as the development of the brain in adolescence can affect decision-making, including long-term planning and the appreciation of abstract concepts such as future health. All information given should be age-appropriate and should be a continuing conversation as the person matures and their needs change. This was highlighted by NCEPOD in 'The Inbetweeners' a review of the transition from child into adult healthcare services.[4]

Patient population

All children and young adults aged 0-24 years, coded for a diagnosis of JIA*, who were being seen by the rheumatology department as an outpatient or who presented to hospital between 1st April 2021 and 31st March 2023, were identified. From this larger group, patients were randomly selected for the peer review if their diagnosis had been made between 1st April 2019 and 31st March 2023, and they were diagnosed or experienced symptoms before their 16th birthday. Due to this sampling method patients aged 20 years and older were not included in the peer review process.
**ICD10 codes: L40.54, M08.0 to M08.9, M09.0, M09.8 and SNOMED codes if required*

Exclusion criteria
Juvenile arthritis in Crohn's disease (regional enteritis) and juvenile arthritis in ulcerative colitis.

Clinical and organisational issues

- Not all rheumatology services were specially commissioned, with adolescent rheumatology being less frequently commissioned (paediatric 33/52; adolescent 16/41; adult 32/45).
- Clinical recognition of JIA was identified as a barrier to rapid referral to rheumatology. Few 23/101 (22.8%) GP practices reported having protocols for the investigation and care of patients with suspected JIA and, when consulting their GP, 20/54 parents/carers felt that they were not taken seriously.
- Variability in care pathways resulted in delays in assessment by a rheumatologist and diagnosis. Only 12/58 patients were referred directly to a rheumatologist and 71/266 (26.7%) patients had a delay in assessment by a rheumatologist. The reviewers believed that diagnosis was delayed in 93/274 (33.9%) patients.
- Patients and parent/carers were not always appropriately trained to administer medication. 22/118 (18.6%) patients and parents/carers had no evidence of being trained in how to give methotrexate injections. Inappropriate medications were given to 26/298 (8.7%) patients while they waited for training on how to administer subcutaneous methotrexate.
- The most delayed medications were intra-articular steroid injections (46/185; 24.9%), subcutaneous methotrexate (32/158; 20.3%) and subcutaneous biologics (23/124; 18.5%).
- A holistic approach to the care of patients diagnosed with JIA was lacking for a significant number of patients. Under half (114/262; 43.5%) of patients had advice and information to support their holistic health. 134/154 (87.0%) clinicians identified gaps in staffing. The most common gaps were in the provision of clinical nurse specialists, psychology, physiotherapy and occupational therapy.

Key features of a service

1. Ensure there are clear and publicised lines of referral pathways to the service with measurable timelines for patients with suspected juvenile idiopathic arthritis.
Clearer lines of referral are needed to ensure that treatment starts promptly and that all necessary multidisciplinary input is arranged. Discussion amongst the clinical groups involved in the study showed that getting referred to rheumatology quickly was often based on luck, with many clinicians reflecting on how parents had to advocate for their child based on their own research or after multiple visits to their GP.

2. Provide timely access to appropriately trained physiotherapy, occupational therapy, pain and psychology services at diagnosis and on an ongoing basis
There was a decline in access to these services as the young person moved to adulthood, however many

patients have JIA as adults and so equivalent access to care needs to continue. Gaps in the service the hospital provides for patients with JIA need to be identified and metrics developed to optimise staffing. Guidance should be established on standardising the multidisciplinary team in terms of which clinicians should be included and how often each patient should be discussed and assessed.

3. Age-appropriate information about JIA and the risks and benefits of medication needs to be provided at diagnosis and on an ongoing basis

Improving understanding and empowering patients and their carers to be involved in making informed decisions about their management will reduce unnecessary delays in starting a treatment due to patient/carer concerns and improve subsequent adherence and ensure treatment starts promptly and continues effectively. Ongoing education and training should be accessible to all patients and carers, and provided in developmentally appropriate formats. Both online and physical resources are still very important to patients and families.

Departments could signpost patients and carers to appropriate online resources, the latest research findings, and JIA support groups to ensure that they are visible to patients and their parents/carers. Parents/carers should be given a contact in the service should they have any concerns or questions about medication. Developmentally appropriate resources for the young person could also be made available around disease therapy, peer support and self-management. Regular re-education could be given as the young person gets older and reaches different life points.

4. Provide training, if age-appropriate, on how to administer subcutaneous injections for JIA at the point treatment is initiated

There were delays to treatment starting as lack of training of how to administer injections meant the medication could not be administered. Training should be undertaken at the time of prescribing. Instruction sheets and videos in English and other languages relevant to your population, including British Sign Language, will help give patients and parent/carers more confidence in administering injections at home.

5. Ensure timely access to intra-articular steroid injections by staff who have been trained to deliver age-appropriate care in units where local or general anaesthesia can be delivered.

Access to medical treatments and home care service should be equitable and not subject to social determinants of health or distance to travel to appointments. Often patients needing intra-articular joint injections required a general anaesthetic but could not access theatre lists.

This could work well when a relationship is formed between rheumatology/surgery/anaesthesia to allow a slot to be made available on a regular list, recognising the ad hoc nature of this patient group needing a ‘medically’ invasive procedure. It may be difficult to fill a traditional list on a regular basis with joint injections, so flexibility is required - possibly bookable semi-urgent slots.

6. Provide a holistic, developmentally appropriate rheumatology service for patients with juvenile idiopathic arthritis

High quality youth work, self-management opportunities and family support can make a significant difference to many young people and their families, especially at diagnosis, during flare ups, and at the time of transition to adult care. Rheumatology provision can be made more patient centred by:

- Considering co-production of the service with young people
- Holding clinics outside of school/college hours
- Offering online appointments and patient/parent/carer access to electronic medical records.
- Providing opportunities for adolescent patients to be seen alone
- Using ‘apps’ and text messaging to inform patients about JIA, to allow them to monitor their symptoms
- Running joint clinics with the paediatric and adult rheumatology teams; members of both teams being present for at least one visit prior to transfer. Other professionals from the wider MDT who

understand or have additional training around the needs of adolescents (not just paediatrics or adult healthcare) and follow adolescent best practice such as those outlined by BANNAR Network Adolescent Care Top Tips should be involved
- Making sure appointments are long enough to cover all aspects of healthcare, and specifically the transition between child and adult services - see 'The Inbetweeners' report

Community diagnostic centres would be an ideal place for paediatric rheumatology teams to deliver health education and potentially peer support opportunities for patients and for parents/carers.

Supporting documents

- **National Rheumatoid Arthritis Service: Juvenile Idiopathic Arthritis**
- **NHS England: Clinical Commissioning Policy Statement: Biologic Therapies for the treatment of Juvenile Idiopathic Arthritis (JIA)**
- **National Institute for Health and Care Excellence: Technology appraisal guidance TA735. Tofacitinib for treating juvenile idiopathic arthritis.**
- **National Confidential Enquiry into Patient Outcome and Death (NCEPOD): The Inbetweeners. A review of the barriers and facilitators in the process of the transition of children and young people with complex chronic health conditions moving into adult health services**
- **Versus Arthritis: Juvenile Idiopathic Arthritis**
- **Arthritis and Musculoskeletal Alliance (ARMA) and the British Society for Paediatric and Adolescent Rheumatology (BSPAR): Standards of care for children and young people with Juvenile Idiopathic Arthritis**
- **British Society for Paediatric and Adolescent Rheumatology (BSPAR) and Royal College of Ophthalmologists: Guidelines for Screening for Uveitis in Juvenile Idiopathic Arthritis (JIA)**
- **National Institute for Health and Care Excellence: NICE Guideline 43. Transition from children's to adults' services for young people using health or social care services**
- **TeachMe Paediatrics: HEADSSS Assessment**
- **National Voices: Ask How I Am**