Review

What does an adult rheumatologist need to know about juvenile idiopathic arthritis?

Elizabeth J. Coulson¹, Helen J. M. Hanson¹ and Helen E. Foster¹,²

Abstract

JIA is the most common chronic inflammatory arthritis in children and young people and an estimated one-third of individuals will have persistent active disease into adulthood. There are a number of key differences in the clinical manifestations, assessment and management of JIA compared with adult-onset arthritis. Transition and transfer to adult services present significant challenges for many patients, their families and health care professionals. We describe key clinical issues relevant to adult rheumatology health care teams responsible for ongoing care of these young people.

Key words: adult juvenile idiopathic arthritis, transitional care, biologics, co-morbidity, multidisciplinary care, outcomes.

Introduction

JIA describes a clinically heterogeneous group of chronic arthritides with age of onset under 16 years of age, an incidence of 1 in 10 000 and a prevalence of 0.1% [1, 2]. Many individuals (an estimated one-third) have persistent active disease into adulthood requiring immunosuppressive treatment [3, 4]. It is essential that the adult rheumatology multidisciplinary team (MDT) have the necessary knowledge and skills to engage with paediatric rheumatology services to deliver transitional care which are integral to the British Society for Paediatric and Adolescent Rheumatology (BSPAR) Standards of Care in JIA [5]. This article focuses on the adolescent and young adult (AYA) with JIA and highlights key clinical issues that the adult rheumatologist may encounter.

Classification of JIA

The ILAR [6] classification for JIA (Table 1) reflects disease heterogeneity and replaces previous nomenclatures (JRA and JCA). The JIA subtype is defined by disease course in the first 6 months from onset and is a helpful prognostic indicator, although there are no studies to validate the ILAR criteria into adulthood and outcome data are largely based on studies reflecting treatment approaches from decades ago. It is envisaged that ongoing inception cohort studies [7, 8] will have extended follow-up into adulthood and in due course will report improving outcomes. Despite these limitations, knowledge of JIA subtypes and their clinical course is important to appreciate differences from adult-onset arthritides.

Systemic-onset JIA

Systemic-onset JIA (SOJIA) is rare (4–17% of JIA) with similar clinical features to adult-onset Still’s disease [classically with fever (one or two spikes daily, usually evenings), rash (salmon pink, evanescent, macular) and arthritis (often polyarthritis)], high acute phase reactants, hyperferritinaemia and sometimes with serositis, hepatosplenomegaly or lymphadenopathy [9]. The pattern of polyarthritis is variable and may not present until several weeks into the disease course, but with a predilection for the hip, ankle, wrist and temporomandibular (TMJ) joints. SOJIA has significant morbidity and potential mortality from macrophage activation syndrome [MAS (see below)]. Amyloidosis, which was a previously observed and often fatal complication in SOJIA, is now rarely encountered [10], but it should be considered in adult patients with proteinuria, renal dysfunction or hypertension and who predate MTX or biologic therapies.

Oligoarticular JIA

Oligoarticular JIA (four or fewer joints affected within the first 6 months) is further categorized into persistent (restricted to four or fewer joints) or extended oligoarticular JIA (more than four joints beyond the first 6 months).
Oligoarticular JIA typically presents in very young children, usually with lower limb involvement (knee or ankle) [1]. These children have a better prognosis with regard to joint disease but have a higher risk of chronic anterior uveitis, especially young girls within the first year of disease onset and those who carry ANA. In contrast to the acutely painful red eye in adults with HLA-B27-associated anterior uveitis, JIA-associated uveitis is classically silent, asymptomatic and, if not detected and treated early, carries a high risk of visual impairment and ocular complications [11]. The activity and severity of uveitis and arthritis do not correlate [12] and all children with suspected JIA need regular screening with slit lamp examination by an ophthalmologist. The recommendations for eye screening vary by perceived risk (with most regular screening in young females with recent oligoarticular JIA who carry ANA) and usually can cease if there has been no uveitis detected after 5 years of follow-up or patients reach 12 years of age [5].

Extended oligoarticular JIA, occurs in one-third of children with oligoarthritis and is usually an asymmetrical polyarthritis with potential for early joint damage, warranting systemic immunosuppression. Predicting extended disease is a challenge, but it is more likely in those children with upper limb involvement and high ESR at presentation [13].

Polyarticular JIA

Polyarticular JIA refers to five or more joints affected within the first 6 months of onset, is usually symmetrical and is further divided by the absence or presence of RF IgM. RF-negative polyarticular JIA is common (30–40% of JIA), associates with uveitis, especially in those with ANA, and the differential diagnosis needs to consider SLE. RF-positive polyarticular JIA is rare (2–7% of JIA), more likely in older children (often teenage females) and uveitis is uncommon [14]; similar to adult RA, anti-CCP antibodies may be present with concomitant risk of worse disease activity and course [15].

Psoriatic arthritis

PsA is rare in children but is likely underdiagnosed, as the rash may appear years after the onset of joint disease. Joint involvement is variable, but is typically asymmetrical, involving large and small joints with dactylitis. Uveitis can occur and can be severe. Distinguishing oligoarticular JIA from juvenile PsA (JPsA) with an oligoarticular pattern is difficult in the absence of rash, but it is important since JPsA has a worse prognosis; JPsA is suggested by asymmetrical, small joint, especially upper limb, involvement and a family history of psoriasis in a first-degree relative [16].

Enthesitis-related arthritis

Enthesitis-related arthritis (ERA) is characterized by enthesitis and arthritis or arthritis and more than two of the following: sacroiliac joint tenderness and/or inflammatory lumbar pain, HLA-B27 positivity, onset in a male >6 years old, acute anterior uveitis or a first-degree relative with SpA (Table 1). Lower limb involvement is common, often asymmetrical, and can follow an aggressive disease course, with those who carry HLA-B27 being more likely to develop sacroiliitis in teenage years or young adulthood. Anterior uveitis is common, typically an acute

---

**Table 1** The ILAR classification of JIA

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Clinical features</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at onset</td>
<td>&lt;16 years</td>
</tr>
<tr>
<td>Minimum duration</td>
<td>6 weeks</td>
</tr>
<tr>
<td>Subtypes</td>
<td>Arthritis</td>
</tr>
<tr>
<td>Systemic</td>
<td>Fever, rash</td>
</tr>
<tr>
<td>Oligoarthritis</td>
<td>Up to four joints affected during the first 6 months</td>
</tr>
<tr>
<td>Polyarthritis</td>
<td>Persistent—affects no more than four joints throughout its course</td>
</tr>
<tr>
<td></td>
<td>Extended—affects five or more joints after the first 6 months</td>
</tr>
<tr>
<td>PsA</td>
<td>Arthritis and enthesitis, or arthritis or enthesitis with at least two of the following:</td>
</tr>
<tr>
<td></td>
<td>Sacroiliac joint tenderness</td>
</tr>
<tr>
<td></td>
<td>Inflammatory back pain</td>
</tr>
<tr>
<td></td>
<td>HLA-B27 positive</td>
</tr>
<tr>
<td></td>
<td>Family history of SpA- or HLA-B27-positive-related disease</td>
</tr>
<tr>
<td>Undifferentiated</td>
<td>Arthritis of unknown cause or not fulfilling the above categories</td>
</tr>
</tbody>
</table>
red eye, in contrast to the chronic anterior uveitis in young children with oligoarticular JIA. Therefore children with ERA undergo less frequent eye screening since they are likely to present with symptoms.

Undifferentiated JIA

Undifferentiated JIA refers to overlap between subtypes (e.g. RF-positive polyarthritis with psoriasis) and the course is variable. The JIA subtypes observed in adult clinics differ from those of paediatric practice. In paediatric rheumatology clinics, oligoarticular JIA is most common (two-thirds of cases), followed by RF-negative polyarthritis and other subtypes are less common [1]. In adult JIA practice, most patients have polyarticular disease irrespective of the subtype at onset, including those with SOJIA, RF-positive and RF-negative polyarthritis, extended oligoarticular JIA, JPsA and ERA [17–19]. Persistent oligoarticular JIA is less common in adult practice, as many children do well and are discharged; exceptions are those with uveitis or intermittent flares [14, 19]. De novo flares of oligoarthritis do occur in adulthood, often after years of remission and typically post-partum; adults with inflammatory sacroiliitis may have a previous history of oligoarticular JIA or ERA and physical signs may be apparent (e.g. loss of hyperextension at a knee or leg length discrepancy from previous monoarthritis).

Management of JIA

Specialist paediatric rheumatology MDTs are integral to optimal management with exponentially increasing use of IA corticosteroids, DMARDs, biologic therapies and rapid escalation of treatment to induce early remission [1, 17, 20–22]. Complications such as growth retardation and localized growth abnormalities (e.g. leg length inequality, micrognathia) are now much less commonly observed unless there has been a delay in access to appropriate care. Adult JIA clinics that include older patients who predate the use of MTX (early 1980s) illustrate the serious consequences of untreated JIA. It is essential that these clinics be included as part of both adult and paediatric rheumatology training and highlight the impact of potent therapies now available.

Recent North American guidelines recommend DMARDs for children with JIA if disease control is not achieved with NSAIDS or joint injections [23]. MTX, the DMARD of choice, is introduced early for all JIA subtypes with the exception of oligoarticular JIA (although involvement of critical joints such as the wrist, TMJ or hip will invariably prompt MTX introduction). MTX is deemed to be safe and efficacious, avoiding long-term systemic steroids and reducing joint damage [24, 25]. MTX is increasingly administered subcutaneously to improve bioavailability, although nausea limits its use for many children and young people. MTX is less likely to control systemic features of SOJIA [24] but is recommended for arthritis [23]. SSZ is effective in HLA-B27-positive oligoarticular JIA [26] and recommended in ERA [23], but it should be avoided in active SOJIA in view of its potential to induce MAS [27, 28]. LEF is effective in polyarticular JIA, with similar tolerability to MTX [29, 30], although implications for planning pregnancy are important for young females [31]. HCQ, penicillamine and gold are rarely used following studies showing little benefit over placebo [32–36], but they are sometimes used if MTX or biologic drugs are not available. In adults with JIA, MTX is frequently used (often subcutaneously), and anecdotally in combination with one or more DMARDs (such as SSZ, LEF or HCQ), albeit without published efficacy data. Ciclosporin may control systemic features in SOJIA and MAS [37–39], although other agents, including cytotoxics or biologics, are increasingly being used.

IA steroid injections are useful in the management of disease flares as bridging measures while starting DMARDs or escalating systemic immunosuppression and may be used as monotherapy in oligoarthritis [23]. Triamcinolone hexacetonide is more efficacious than triamcinolone acetonide based on studies in children [40, 41]. General anaesthesia is needed for young children, multiple joint injections or less accessible joints (e.g. TMJ, hip) and imaging (especially US) is increasingly being used to improve accuracy. Inhaled anaesthesia for joint injections [42] is advocated [5], provides safe and rapid access to treatment [43] and is well tolerated in older children.

A review of biologic drugs for JIA in children and young people is beyond the scope of this article and reviews are suggested [44–46]. Etanercept was the first anti-TNF to be licensed in children (aged 2–17 years) with JIA [47] and other biologics (adalimumab, abatacept and tocilizumab) have been licensed for use. In the UK, etanercept is National Institute for Health and Care Excellence (NICE) approved for children aged 4–17 years with JIA with five or more active joints unresponsive to or intolerant of MTX [48]; this contrasts with adult RA guidance where patients are required to fail two DMARDs [49]. Tocilizumab is NICE approved as a first-line biologic for active SOJIA unresponsive to or intolerant of MTX [50] and, notably, guidance does not include an upper age limit. National registries demonstrate the use of etanercept and other biologics in all JIA subtypes [51–56] and studies demonstrate the efficacy of etanercept as monotherapy or in combination with MTX in polyarticular JIA, JPsA and ERA as well as in early disease [57–60].

Many children and young people with JIA (at least one-third [61, 62]) are likely to need ongoing immunosuppressive treatment into adulthood, and persistent disease activity despite DMARDs is not uncommon. The British Society for Rheumatology Biologics Registry (BSRBR) for adult RA reports biologic therapies are often used and off-license in adults with JIA [53]. Etanercept is used most frequently (49%), followed by infliximab (28%), adalimumab (22%), anakinra (1%) and rituximab (<1%). Furthermore, BSRBR data show that many (50%) adults with JIA received more than one biologic during the study period, with 22% receiving more than three anti-TNF therapies and inefficacy being the most cited reason for class switch [53].
There are no clinical trials of biologics in adult JIA to demonstrate efficacy or inform choice of drug or response to treatment. The Juvenile Arthritis Disease Activity Score (JADAS) and JADAS 3 [63, 64] offer objective feasible measures to facilitate a treat-to-target approach in JIA [65], but they need further validation and have not been used in adults. Furthermore, there is no guidance for sequential biologics or their use in isolated sacroiliitis or uveitis. Anecdotally the age restriction for some licensed biologics and the lack of NICE guidance for adults with JIA has resulted in inequity in access to biologic therapies in the UK. In response, the combined British Society for Rheumatology (BSR)/BSPAR statement [66] recommends that ongoing treatment with biologics be based on clinical need, irrespective of age, and that consensus about adult JIA is urgently needed. NICE has agreed to review the guidance for the use of biologics in JIA and extend this guidance to adults with JIA. This is welcome, albeit unlikely to be available until at least 2015.

**Key clinical issues**

We consider clinical issues frequently encountered and key points are summarized in Table 2.

**Disease flares**

Flares of disease may occur as a *de novo* presentation in adulthood, often after years of remission and may be the first presentation to adult services. There is considerable variation in the clinical presentations of disease flare and these reflect the spectrum of JIA subtypes. Monoarthritis flares in oligoarticular JIA, JPsA or ERA can occur and polyarticular flares can occur with all subtypes. It is important to remember that acute phase reactants can be normal in active JIA and that ANA or RF can persist based on clinical need, irrespective of age, and that consensus about adult JIA is urgently needed. NICE has agreed to review the guidance for the use of biologics in JIA and extend this guidance to adults with JIA. This is welcome, albeit unlikely to be available until at least 2015.

**Biologic therapies**

In the absence of consensus guidance or clinical trials in adult JIA, the choice of biologic therapy depends on patient choice, clinician experience, adherence factors, the presence of uveitis and the JIA subtype with extrapolation from paediatric studies in JIA or clinical trials in adult inflammatory arthritis. We present our opinion based on experience, informed by paediatric practice and clinical studies in adult-onset arthritides.

Etanercept and adalimumab may be used as monotherapy for adults who are intolerant of MTX or weaning off treatment prior to pregnancy planning [23]. Adalimumab and infliximab are more likely to be used if there is a history of uveitis [71–73]. Infliximab is a useful option where adherence is suboptimal. Despite previous intolerance of high-dose MTX, many adults may tolerate low-dose MTX in combination with infliximab or another biologic to optimize efficacy and avoid neutralizing antibodies [58, 74]. Individuals with RF-positive polyarticular disease respond well to rituximab [58, 74]. For MTX-resistant polyarthritis, irrespective of onset subtype, abatacept has shown benefit [75]. Switching to a second anti-TNF is common and may be effective in patients with extended oligoarticular JIA, RF-positive or -negative polyarticular JIA and ERA subtypes [54, 76]. Options for managing SOJIA depend on the presence or absence of active systemic features, arthritis or both. IL-1 blockade appears effective for active SOJIA with systemic features, but less so for articular disease [23]. Based on recent paediatric studies, tocilizumab is useful to control systemic features or polyarthritis in SOJIA [77] and is effective in polyarticular JIA [78].

It is important to counsel patients and document the efficacy of biologic therapies. The role of registries to establish the safety and efficacy of biologics is important and hopefully aided by recent initiatives to facilitate shared datasets and long-term follow-up [79]. The potential risk of malignancy remains controversial and recent studies suggest that JIA per se, irrespective of DMARDS or biologics, may be associated with increased malignancy [80]. Fertility and pregnancy outcomes for patients exposed to biologics appear good, although follow-up into adulthood is very limited for those who started treatment as children.

When to stop biologic treatments is an important issue, although there is a lack of guidance or evidence to inform decision-making. Our advice is that cessation of treatment is reasonable after 2 years of clinical remission, but there is the risk of flare and it is important to plan a trial off-treatment with the patient, taking into account life events (such as pregnancy planning, work or higher education opportunities).

**Uveitis**

Silent uveitis developing *de novo* in adults with JIA is rare [81] and routine eye screening is not regular clinical practice for adult JIA. However, grumbling chronic anterior uveitis can continue [81] and may flare with changes in treatment (such as cessation of treatment...
**Table 2** Key clinical issues in the management of adults with JIA

### Disease flares
- Consider macrophage activation in unwell patients with JIA.
- Laboratory markers can be normal in disease flares.
- Consider sepsis in a single joint flare.
- Consider inhaled anaesthesia for joint aspiration and steroid injections.

### DMARDs and biologics

<table>
<thead>
<tr>
<th>Disease subtype</th>
<th>DMARDs</th>
<th>Biologic therapy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Polyarticular course JIA</td>
<td>MTX, SSZ, LEF, HCQ</td>
<td>Anti-TNF; abatacept/rituximab/tocilizumab</td>
</tr>
</tbody>
</table>

### Uveitis
- Uveitis is often silent and may continue into adulthood. Flares can occur in adulthood, especially during breaks in treatment or switching agents. May not flare coexistent with arthritis activity.
- Seek more regular ophthalmology review if:
  - Tapering biologics or DMARD treatments
  - Stopping biologics or DMARD treatments (e.g. for planned pregnancy)
  - Changing biologics or DMARD treatment regime
- Caution in monotherapy with etanercept.

### Vaccinations
- Check measles, rubella and varicella serology and advise patient and health care providers accordingly.
- Be alert to windows of opportunity for immunisation catch-up of live vaccines: e.g. breaks in treatment prior to planned pregnancy.
- Advise patient to have influenza and pneumococcal vaccination.
- Seek advice for live vaccinations that may be needed and avoid if taking biologics, corticosteroids or combination with DMARDS.

### Temporomandibular joint disease and oral health
- Generic health advice includes good oral hygiene and dental reviews.

### Cardiovascular health
- Assessment of cardiovascular risk factors.

### Pregnancy
- Discontinue biologics and DMARDs as per guidance in adult rheumatology.
- In patients with severe disease, consider the risks and benefits of continuation of anti-TNF therapy and discuss with shared care obstetrician.
- Counsel that significant hip involvement or short stature may necessitate caesarean section.
- Be alert to flares of chronic anterior uveitis with breaks in treatment.

### Bone health
- Perform DXA at baseline and subsequently every 2 years.
- Advise patient to use calcium and vitamin D supplementation.
- Advise patient to perform weight-bearing exercise.
- Avoid smoking and alcohol excess.
- In women, use combined oral contraception.

### Transitional care
- Adult rheumatology MDT needs to have skills and training in JIA and adolescent health.
- Transitional care starts in early adolescence; adult rheumatology teams liaise with paediatric rheumatology to facilitate joined up working and communication.
- Models of transitional care reflecting local expertise and clinical networks.
- Adult rheumatology to include transitional care and adolescent health as an integral part of training.

SOJIA: systemic onset JIA; MDT: multidisciplinary team.
prior to pregnancy) and shared care with an ophthalmologist experienced in JIA-associated uveitis is advocated. To date, there are no published clinical trials of biologics in JIA uveitis, and clinical practice is influenced by case reports. Etanercept appears not to improve outcomes in uveitis [82, 83], and new development of uveitis or flares may occur while on monotherapy [71]. Adalimumab and infliximab are deemed effective and are used preferentially if there is history of ocular disease [71–73]. Tocilizumab [84], rituximab [85] and abatacept [86] may also be effective.

Vaccinations

Patients with JIA may have incomplete immunization status [87, 88]. It is worth checking status for measles and rubella as well as varicella zoster virus (VZV) for all adult JIA patients, with advice given accordingly. Acute de novo infection in the young adult, especially VZV or measles, can be life-threatening. In the UK, where VZV vaccination is not part of the national immunization programme [89], children who are VZV non-immune are offered VZV vaccination before they start DMARDs if there is an appropriate window of opportunity. The European League Against Rheumatism (EULAR) states that non-live vaccines appear to be safe and effective in children with JIA, given according to national vaccination guidelines and prior to rituximab to optimize the immune response [90]. Similar advice would seem appropriate for adults with JIA. The EULAR also suggests that certain live vaccines may be administered safely and effectively in patients on low-dose MTX as monotherapy [90], though caution is warranted, and for patients taking combination immunosuppressives, including biologics, MTX and corticosteroids, live vaccines are not recommended and expert opinion should be sought in the presence of exposure to infection (e.g. measles). Breaks in treatment (due to disease remission or planned pregnancy) may allow catch-up for live immunizations (e.g. measles, mumps and rubella and VZV). For young adults, especially those planning to travel to endemic areas, yellow fever vaccination may also be appropriate.

Pregnancy and contraception

Outcomes for pregnancy in JIA appear overall to be good, with a reduction in disease activity often occurring during pregnancy but with an increased risk of post-partum flare [92]. It is unclear what impact current treatment strategies have on pregnancy outcomes, but anecdotaly many adults with JIA deliver healthy babies. The published literature reflects previous treatment strategies and is subsequently biased to include patients with complications of prolonged disease activity (such as short stature and hip disease); studies show higher rates of pre-eclampsia and post-partum haemorrhage but no clear increase in adverse neonatal outcomes [93]. Pelvic inflammatory disease, ovarian cysts and premature ovarian failure may be more common [94, 95].

Current advice is that planning ahead before conception is important, with discontinuation of MTX, other DMARDs and biologic therapies according to guidelines in adult arthritis [31, 96]. Potential flares in disease activity (especially uveitis) and the impact of short stature or hip disease need to be considered; ideally the patient should receive shared care with obstetrics and ophthalmology as appropriate.

Generic sexual health and contraception advice is similar for adults with RA [97]; specialist advice may be required for those contemplating intrauterine devices and if taking biologic therapies.

Bone health

Risk factors for osteopenia in JIA include active disease, delayed puberty, reduced physical activity, nutritional deficiency and corticosteroid use [98]. Limited data are available in AYA patients with JIA, but low BMD and increased fracture rates associated with childhood arthritis extend into adulthood [99–101], particularly in those with active disease [102].

Assessing bone density is challenging; typically BMD accelerates in puberty and peaks between 25 and 35 years of age [103]. DXA is widely used to assess BMD in older adults and correlates with future fracture risk [104]. The relationship between DXA-determined BMD and fracture risk in healthy AYAs is unclear [105]: a low T score in an AYA may reflect skeletal size or delayed puberty due to co-existing chronic illness.

Lifestyle changes (weight-bearing exercise, avoidance of smoking/alcohol excess and promotion of dietary calcium intake) as well as optimizing disease control and low exposure to corticosteroids are important. Baseline BMD and subsequent measurement every 2 years is advocated in adult patients [101]. In those with abnormal BMD, calcium and vitamin D supplementation and in females the use of combined oral contraceptives is advocated, but bisphosphonates are not licensed for premenopausal women [106]. Our practice is that bisphosphonates are restricted to males with low-trauma fractures and post-child-bearing females, with specialist advice recommended.

TMJ disease and oral health

TMJ disease in prepubertal children with JIA can result in malocclusion and micrognathia [107, 108], but it is often overlooked [109]. Poor oral health and higher rates of caries are reported in JIA at all ages [110], and TMJ disease is likely to be contributory. Good oral hygiene is advocated, especially for those on immunosuppressive treatment, due to the risk of bacteraeemia. Established TMJ disease may need splinting [111], but optimal disease control to avoid joint damage and early referral to orthodontic services are advocated [112, 113].

Cardiovascular health

The EULAR recommends annual cardiovascular risk assessment in adult-onset arthritides [114]. Preliminary studies also suggest an increased risk of cardiovascular
disease in JIA, and assessment of cardiovascular risk factors should be considered as part of routine clinical care in adults with JIA [115].

Orthopaedic intervention and operative risk
The need for joint replacement surgery is decreasing and may reflect changes in treatment strategies [116, 117]; nonetheless, some adults with JIA may need orthopaedic opinion and synovectomy (or surgical) for refractory single-joint disease [118]. With any procedure involving general anaesthesia, cervical spine and specialist anaesthetic assessment may be warranted. Invasive procedures may require temporary cessation of biologic therapies and antibiotic prophylaxis pending the procedure.

Transitional care
Transition is defined as a multifaceted active process that attends to the medical, psychosocial and educational/vocational needs of adolescents as they move from child- to adult-centred care [119]. Transfer to adult rheumatology may involve the change to a different hospital, potentially in a different town or city, all of which may be unfamiliar to the young person. Transfer without appropriate preparation (as part of transitional care) or at a time that coincides with challenging circumstances, such as ill health, examinations or family/social problems, has been shown to associate with poor clinical attendance, lower rates of contact, reduced adherence and worse clinical outcomes [120–122]. Adolescence involves consolidating identity, establishing relationships outside the family, achieving independence and finding a meaningful vocation [123]. Many AYAs have not achieved these goals when they arrive in adult care [124].

Transitional care is a complex package of care [125] that starts early (usually from the age of 11 years, incorporates the transfer (usually at 16–19 years of age) and extends into the young adult years (~25 years of age). Ideally all young people will have a planned, coordinated transition and transfer to an adult rheumatology MDT with experience in managing adult JIA [5]. The MDT needs to have the skills, training and resources to support young people, provide referrals to other agencies [125, 126] and acknowledge the importance of following up on non-attendance to safeguard the patient.

Transitional care addresses generic and mental health [HEADSSS (home, education, activities, drugs, sex, suicide, sleep)] [127] as well as disease-specific concerns (e.g. regarding MTX with advice about alcohol, contraception and sexual health). Adherence is key to optimal disease control; non-adherence may reflect poor understanding of the prescribed medication and its impact (perceived or real) and side effects (such as nausea with MTX) or it may reflect a desire to be involved in a shared decision-making process.

Many AYAs with JIA transferring to adult care can expect to have good disease control, although they are likely to be taking complex treatment regimes, including biologics and DMARDs. Models of transitional care are variable, depending on local services and the availability of appropriately trained health care providers. In the UK, the Department of Health has quality criteria for providing adolescent-centred health services [128]; key themes include the accessibility of services, staff training, encouraging shared decision-making and ensuring confidentiality.

Employment and work stability are key outcomes of a successful transition [129, 130] and these can be improved through part-time working, work experience, hobbies and active discussion about careers—ideally to mitigate the expectation of disability and unemployment based on outcomes studies [61, 62, 131, 132]. Chronic illness may impact on vocational readiness, and health care teams are likely to play an important role in supporting AYAs with disclosure to employers, peers or tutors [133].

Summary
Adult rheumatologists need to be aware of the wide range of issues, including transitional care, work and employment, sexual health and pregnancy and potential long-term co-morbidities including bone health and cardiovascular disease. There is no consensus for the use of DMARDs and biologic therapies in adults with JIA, with management based on extrapolation from paediatric rheumatology or adult practice. Careful counselling and documentation is required to address the many uncertainties about treatment options and long-term safety. Education and training for adult MDTs involved in adult JIA care is strongly advocated and suggested resources include http://www.unil.ch/euteach, http://www.e-lfh.org.uk, http://www.bspar.org.uk/adolescent-rheumatology and http://www.yphsig.org.uk/training.

Rheumatology key messages
- Ongoing care for adults with JIA should be delivered by adult rheumatology multidisciplinary teams with appropriate skills and experience.
- Treatment strategies are often based on extrapolation from paediatric practice and clinical studies of adult RA.
- Further guidance for treatment of adult JIA is needed.

Acknowledgements
Dr Coulson’s salary is funded by the National Institute for Health Research (NIHR) Clinical Research Network (CRN) and the Tyne and Wear and Northumberland Community Foundation (Baines Fund).

Disclosure statement: H.E.F. has received consultation honoraria, educational grants and bursaries from Pfizer, AbbVie, Roche, Novartis and Schering Plough. H.H. is currently employed on a grant supported by Pfizer. E.C. has declared no conflicts of interest.
References


