LESSON OF THE MONTH

Systemic juvenile idiopathic arthritis presenting in a young child with long term disability as an adolescent

K M Bailey, J E McDonagh, A M Prieur

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CASE HISTORY

A young man now aged 19 years presented, aged 2½ years, with an 8 week history of being unwell with high swinging fevers, a ''measles''-type rash, anorexia, and weight loss. He complained throughout this time of a painful right knee, and subsequently, pain in his left wrist and ankle, neck, and elbows. Initial assessment confirmed synovitis in his left ankle, right hip, neck, and some small joints of his hands. He had a quotidian fever pattern, intermittent macular rash, lymphadenopathy but no splenomegaly, and a cardiovascular examination was normal. Inflammatory markers were high (erythrocyte sedimentation rate (ESR) 129 mm/1st h; C reactive protein (CRP) 226 mg/l; platelets 594×10^9/l—peak 1360×10^9/l), haemoglobin 92 g/l; white blood cell count 9.6×10^9/l (neutrophils 6.6, lymphocytes 1.9), and albumin 24 g/l. The differential diagnosis included systemic arthritis, infections, inflammatory bowel disease, and malignancy.

After exclusion of these diseases, a diagnosis of systemic juvenile idiopathic arthritis (SJIA) was made, and treatment was started with high dose non-steroidal anti-inflammatory drugs and oral prednisolone. Many treatments were subsequently used in the following years, including penicillamine, rifampicin with isoniazid, methotrexate (introduced 4 years into his disease and continued for 7 years), ciclosporin in combination with methotrexate, intravenous immunoglobulin; he is currently receiving etanercept (started after 15 years of disease, maximum dose 0.8 mg/kg twice weekly). These treatments have been complemented with intermittent multiple intra-articular steroids. Despite this treatment he continues to have active disease—ESR 42 mm/1st h, CRP 38 mg/l (ESR <20 mm/1st h on only two occasions in 17 years)—and has remained dependent on steroids (average daily dose 5–10 mg prednisolone).

There have been many consequences of a lifetime of inflammatory disease. Growth restriction (130 cm tall at the time of transfer to adult care, with only a 35 cm gain in height from diagnosis) and delay in puberty have had a major impact in addition to his significant disability. Bullying at school and angry outbursts were significant problems in early to mid-adolescence. Treatment with growth hormone (GH) had minimal effect. Testosterone precipitated puberty at the age of 15–17 years.

Osteoporosis (z score of −3.5) as a consequence of persistent inflammation and extensive corticosteroid treatment has resulted in many fractures, including a spiral fracture to the femur, several vertebral fractures, and he has been treated with intravenous pamidronate. Persistent synovitis has led to extensive joint destruction, disability (Childhood Health Assessment Questionnaire score >2.0), and pain.

He has required multiple operations, including synovectomy and supracondylar osteotomy of both knees. A right total hip replacement at the age of 12 has been problematic and he is likely to require a girdlestone procedure in the future. Both knees have been successfully replaced (age 15 and 17 years), as has the other hip (age 19). Atlantoaxial subluxation, resulting in medullary cord compression, necessitated cervical fusion and odontoidectomy, requiring 9 months in a halo brace at the age of 14 years.

Immune suppression has resulted in severe chicken pox with pneumonitis and subsequent shingles. Methotrexate induced nausea and mouth ulcers, together with apparent lack of efficacy, have led to intermittent withdrawal of treatment and non-adherence. Amyloid studies to date have been normal.

An electric wheelchair allows him some independent mobility and a modified car (and driving lessons), delayed owing to his hip surgery, is now eagerly awaited. He is currently studying at university, and hopes to follow a career in the media, although he has not had any work experience to date. Transfer to the care of an adult rheumatologist occurred when he was aged 19 before starting university during a gap year for his hip surgery. He was initially seen in a combined young adult clinic with his paediatric rheumatologist present.

Learning objectives

- To demonstrate the difference between transition, the process and transfer, the event, for adolescents with juvenile idiopathic arthritis (JIA)
- To recognise the challenges of treatment for continuing inflammatory disease on entry into adulthood
- To recognise reciprocal influences of chronic illness and adolescent development (physical, psychosocial, and cognitive)

DISCUSSION

When is the right time to transfer this young man to adult rheumatology?

There is no arbitrary ''right'' age for the transfer, and timing should be flexible. One of the fundamental lessons of transitional care is the difference between transfer and transition. Transition is most usefully 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''.[1] Transition ideally starts early, usually in early adolescence. Transfer is only one of many events within this process.

Unfortunately, evidence shows that the transfer of such young people with chronic illness is currently poorly handled.[2–4]

Abbreviations: CRP, C reactive protein; ESR, erythrocyte sedimentation rate; GH, growth hormone; JIA, juvenile idiopathic arthritis; IL, interleukin; SJIA, systemic juvenile idiopathic arthritis

Table 1 Determinants of the timing of transfer

Chronological age
Maturity
Completion of growth and puberty
Medical status
Adherence
Independence
Acquisition of skills to function in adult service
Adolescent readiness
Parental readiness
Availability of adult rheumatologist
Timing of other transitions—for example, leaving school

The timing of transfer is multifactorial (table 1) and should be integrated into a coordinated, individualised, transition plan (table 2).

The young man described was prepared for transfer, with discussions documented in early and mid-adolescence, particularly pertinent in view of the severity of his disease. He had also completed his physical growth, was post-pubertal, and was due to start university.

However, he had continuing active disease. Ideally, transfer should take place during a period of remission, but as he had never truly been in remission, a relatively stable phase would be the preferred alternative.

His parents were extremely anxious about leaving the paediatric rheumatology team whom they had known for 17 years and from whom they had received significant support throughout their son's illness. A third of health professionals in a recent nationwide survey reported parental difficulties during transition, and such parental and family factors were perceived to influence the success of transition.[5]

The availability of an adult rheumatologist who is aware of the significant differences between childhood onset arthritis in adulthood and adult onset arthritis is the ideal, particularly for the reciprocal influences of the chronic illness on adolescent growth and development (physical, cognitive, and psychosocial), as exemplified by this patient. The impact of disease on this 19 year old with a 17 year history of JIA differs from that of a 60 year old with a healthy childhood and adolescence and the same disease duration of adult rheumatoid arthritis.

In the young man described, the inflammatory disease and long term steroid treatment had significantly restricted growth and delayed puberty, which in turn had led to bullying at school and behavioural problems in early adolescence. His growth would have also determined the dose regimens of his drugs, the timing of his replacement surgery, and interpretation of dual *x* ray absorptiometry results. Psychosocially he remained dependent on his parents for transport and had to delay starting tertiary education and driving lessons owing to his need for surgery.

Finally, differences in the delivery of care should also be expected—for example, consultation times are significantly shorter in adult rheumatology.[6] The clinical environment is also different, with the average age of patients in an adult clinic significantly older. If the young person is not prepared for these differences, their first experience of adult care may be negative, even ''frightening''.[2 7]

What will be the key components of an individualised transition plan for this young man?

There are three major key components of a transition plan, which, in turn, are interdependent: (*a*) health; (*b*) home; (*c*) vocational. In a nationwide survey, 77% of professionals felt individualised transition plans were ''very important'' for adolescents with JIA.[5] The importance of such plans has also been recognised in several recent major policy statements in the UK and USA.[3 7 8] However, in a nationwide audit of recent transfers to adult rheumatology, only 3% of patients had had a structured transition plan.[9] Blum *et al* reported that lack of planning was second only to finance as a reason for failure of successful transition.[1]

Table 2 Example of an individualised transition plan

Late transition plan (17–19 years) **Transition skills/knowledge**
Health transitions
I am confident in my knowledge about arthritis and its treatment
I understand what is likely to happen with my arthritis when I am an adult
I feel confident to be seen on my own in clinic
I look after my own drugs
I have my joint injections performed without a general anaesthetic
I order and collect my repeat prescriptions and book my own appointments
I call the hospital myself if there is a query about my arthritis and/or treatment
I know the plan for my rheumatology care when I am an adult
I exercise regularly
I understand the risk of sexual behaviour while taking my drugs (if applicable)
I understand the implications of JIA/drug treatment on future child bearing (if applicable)
I know where and how I can access providers of reliable accurate information about sexual health
I understand the effect of smoking, drugs, or alcohol on my arthritis and general health
Home transitions
I am responsible for a particular household chore(s) at home
I am independent at home—dressing, bathing, showering, preparing meals, etc
I can or am learning to drive
I know how to plan ahead for trips/being away from home
I understand my eligibility for benefits (if applicable)
School/vocational transitions
I have a career plan
I have had work/volunteering experience
I am aware of the potential impact (if any) of my arthritis on my future career plans
I know how and what to tell a potential employer about my arthritis (if applicable)
Other transitional care issues (give details)
For each point in the above table the following information should be obtained: 1 Response: Yes/I can do this on my own and/or understand this fully OR I need advice and/or help with this 2 Action plan 3 Key person involved.

Ideally an individualised transition plan would have been developed in early adolescence in consultation with the young person and reviewed regularly, reflecting the course of his physical, cognitive, and psychosocial development through early, mid, and late adolescence. Table 2 describes an example of a transition plan for this patient.

First and foremost, optimal control of his disease is the ideal status for transfer and a key aspect of transition planning and a few aspects of this will now be discussed in detail.

1. Disease-specific issues

- Control of disease activity
- Treatment of related major morbidities
- Treatment of other morbidities
- Potential of future therapeutic trials

Disease activity control

- In a young man with complex disease, for whom all other treatments had failed, maintenance prednisolone

5 mg/day remains an important component of his management to avoid a disease flare.

- ESR and CRP have improved since etanercept was started, suggesting that this drug is efficient in this particular patient. However, in our experience, etanercept is not as efficient in SJIA as in other JIA subtypes.[10] It is therefore likely that this patient will require alternative treatment in the near future (see below).

Treatment of related major morbidities

Growth retardation Body image is closely related to emotional wellbeing in adolescence.[11] Adolescent physical development and hence body image can be affected both by the disease—for example, joint deformity, growth retardation, pubertal delay—and adverse effects of treatment—for example, steroid induced weight gain and/or acne. Evidence from other chronic illnesses suggests that both paediatricians and adult physicians are poor at monitoring growth and pubertal development in chronic illness, and continued attention is required to growth in chronic illness well into the early twenties.[12] In early adolescence this patient unfortunately was bullied as a result of his growth restriction and immobility. Children with short stature are more at risk of bullying than the general population[13] and dealing with bullying has been identified as a component of transitional care by young people with JIA.[2]

In an attempt to improve his growth, the patient received two series of GH administration between 8 and 11 years and 13 and 19 years, respectively. His end height is still extremely low. Several reasons may explain the lack of efficacy of GH: (*a*) the level of inflammation, which seems to be associated with a low efficacy of GH[14 15] and (*b*) the interruption of GH treatment between the ages of 11 and 13, a crucial period for growth. It seems that GH can partly counteract the adverse effects of glucocorticoids on growth and metabolism in patients with chronic inflammatory disease.[16] But the effect of GH on final height may be greatest when it is given early after disease onset and/or at remission.[17]

Pubertal delay, even in healthy children, may cause considerable psychological distress.[18] Some of these issues may have contributed to the angry outbursts of our patient in early adolescence. In a boy, the addition of testosterone, particularly in the prepubertal period can also help improve growth, as occurred in the patient described.

Osteoporosis led to major complications in this patient. Corticosteroids, associated with chronic inflammation and with a reduction of physical activity due to joint involvement, have a catastrophic effect on bone mineral content. Osteoporosis also causes vertebral collapse and avascular necrosis. There is no standardised therapeutic solution. GH treatment can improve bone metabolism.[19] The role of calcium and vitamin D supplementation for the prevention and treatment of osteoporosis associated with paediatric rheumatic diseases remains to be established. New treatments such as bisphosphonates (for example, pamidronate, risedronate, and calcitonin) are now available,[20] and trials in paediatric patients with inflammatory rheumatic disease are eagerly awaited.

Iron deficient anaemia is constant in chronic inflammatory disease. Recently, and of particular relevance to SJIA, interleukin (IL)6 has been identified as a major player in the mechanism of the anaemia of chronic disease.[21] If iron deficiency is confirmed, iron supplementation given intravenously can be helpful.[22] The use of erythropoietin is not logical because these patients have a normal endogenous production of erythropoietin.[23]

Other related morbidities

- Infectious risk. The immunosuppressive drugs used for many years in this patient, render the risk of severe infection very likely. Any infection must be treated thoroughly, and immunosuppressive treatment must be interrupted during that time. Vaccinations with killed antigens must be also given. Some authors give clotrimoxazole to prevent infection.
- Cataracts due to long term corticosteroid treatment should be operated on if visual impairment is a problem. The possibility for lens implantation makes this complication now less difficult to handle in young patients.
- Avascular necrosis and joint damage require joint replacement, as already performed in this patient on knees and hips.

Future therapeutic trials We must be very cautious about the next therapeutic options in this young man. The medical management in adulthood should be as least ''toxic'' as possible, and potentially aggressive drugs should not be given without a hope of efficacy. At this stage of the disease, the risk of iatrogenicity is much higher than the risk of benefit.

There are no controlled trial data currently available on alternative therapeutic options in SJIA. A logical approach should be to target the cytokine which seems to be associated with SJIA—namely, IL6.[24 25] Such drugs are not yet available, but a new biological agent, atlizumab, an anti-IL6 receptor antibody, is at an early stage of evaluation.[26] Some authors have suggested that thalidomide can help disease control in SJIA. The first published data are impressive, but further confirmation is required. The risk of peripheral neuropathy must be verified regularly, as well as that of teratogenicity in sexually active patients.[27] Encouraging results are now available in children treated with autologous bone marrow transplantation.[28] However, the associated mortality risk is still high, and this technique must be applied to patients whose joint destruction remains compatible with a good functional outcome, which is not the case in our patient. Furthermore, owing to the long disease duration, and the long list of immunosuppressive drugs used, the risk of severe infectious complications would render this therapeutic option very hazardous.

After consideration of disease control, other important components of the individualised transition plan include:

- Disease education needs
- Autonomy in health care
- Transfer to adult care.

2. Disease education needs

The young person described here had had his disease since early childhood, and potentially much of the initial disease education may have been primarily directed to the parents. Disease education is an important area to re-examine during transition and before transfer. A significant level of inaccuracy and misunderstanding about their disease has been reported in long term clinic attendees.[29] In a recent study of adolescents with JIA, two thirds did not know what the abbreviation JIA stood for and over a third thought JIA would be known as rheumatoid arthritis when they became 18 years old.[30] Beresford and Sloper also reported that adolescents with a range of chronic illnesses, including JIA, have a considerable need for information, but that considerable barriers exist to these needs being met.[31]

3. Autonomy in health care

The young person described was introduced at age 11 to the idea of being seen by himself and by age 15 years he was independent. Independent visits are important for a variety of reasons, including confidentiality for the young person and skills training for adult centred care. The young person described is somewhat exemplary, however, as data reported from a national audit of patients with JIA recently transferred to adult rheumatology shows that the mean age at the first discussion of transfer is 18 and the mean age at transfer is also 18 years.[9] In recent publications, only 16–27% of teenagers (13+ years) are reported to have been seen independently of their parents in rheumatology clinics.[9 32]

Being seen independently is not so much being seen alone but being able *to choose* who is in the consultation room, a choice considered important by young people with JIA.[2] However, some parents remain anxious about their son/daughter being seen without them. Geenen *et al* reported that parents felt young people should start being seen independently and learning self management skills at a significantly older age than that reported by health professionals.[33] Parents of young people with JIA suggested that this ''transition'' would be made easier for parents if continuity of care was assured and health professionals actively involved the young person in the family consultations in order to demonstrate their ability to conduct the consultation independently.[2]

4. Transfer to adult care

Transfer to adult care was initially discussed when this patient was 15 years of age and he was in one of the nine dedicated adolescent rheumatology clinics in the UK.[34] Definitive transfer eventually took place at age 19 and although he had not met the adult rheumatologist before transfer, he was transferred to the young adult clinic involving his (familiar) adolescent specialist and an adult rheumatologist. Studies in several childhood onset chronic conditions have reported that young people prefer to meet the adult physician before transfer (JIA[2]; cystic fibrosis[35 36]; insulin dependent diabetes mellitus[37]). Various models for transition have been proposed to date but as yet there is no clear evidence as to which components determine successful transition.[38]

Are there training needs within adult rheumatology services which need to be met in order to provide appropriate transitional care for such young people?

In a recent national Delphi study in which users and providers considered the best practice and feasibility of transitional care in the UK health service, providers reported that the feasibility of transitional care being provided by ''professionals who are knowledgeable about adolescent development'' was limited.[39] The variety of unmet education and training needs of healthcare professionals identified in a national service of professionals (including adult rheumatology) supports this finding.[40] Accordingly, a recent recommendation is that ''training in adolescent health should be mandatory for both undergraduates and trainees of all the Royal Colleges whose members may be involved with the care of young people''.[7]

CONCLUSIONS

In summary, this case report exemplifies the challenges facing paediatric and adult rheumatology in the management of patients with severe disease during the transition from child to adult centred care. The need to consider the biological, cognitive, and psychosocial aspects of disease management, all intrinsic to adolescent rheumatology, has been highlighted, as has the significant morbidity of this disease. The increasing recognition of the age- and development-specific needs of adolescents with chronic rheumatic disease such as SJIA is welcomed and further research, service, and training in this area is actively encouraged.

Learning points

- JIA is NOT a benign disease and has significant morbidity (including iatrogenesis) in adulthood.
- Age related complications present challenges in the management of patients with severe SJIA during adolescence.
- The medical management of such patients in adulthood should be as least ''toxic'' as possible and potentially aggressive drugs avoided unless there is realistic hope of efficacy.
- The quality of transitional care is important for young people.
- The impact of chronic illness and growth and development (physical, cognitive, and psychosocial) during adolescence is different from that seen during adulthood.
- Adolescent health and transitional care should be included in training programmes of health professionals who care for such young people, whether in the paediatric or adult sector.

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JEMcD is an Arthritis Research Campaign clinical senior lecturer in paediatric rheumatology (www. arc.org.uk).

Authors' affiliations

K M Bailey, Department of Rheumatology, Birmingham Children's Hospital, Birmingham, UK
J E McDonagh, Institute of Child Health, Birmingham Children's Hospital, Birmingham, UK
A M Prieur, Hopital Des Enfants Malades, Paris, France

Correspondence to: Dr J E McDonagh, Institute of Child Health, Birmingham Children's Hospital and University of Birmingham, Steelhouse Lane, Birmingham B4 6NH, UK; j.e.mcdonagh@bham.ac.uk

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