This review discusses the problems facing adolescents with juvenile idiopathic arthritis (JIA) and how they are managed. ... musculoskeletal conditions, but a detailed discussion concerning these diseases is beyond the scope of this article.

The size and scope of the problem

JIA has a prevalence of approximately 130 per 100,000 children. The classification has recently been changed (Table 1). Although principally a disease of younger children, JIA presents between the ages of 10 and 16 years in 25% of patients. Those who develop systemic or polyarticular disease at a younger age have a higher incidence of destructive joint disease. The common belief is that JIA ‘burns out’ before or during adolescence, so the ensuing problems improve or remain static, but in fact in one-third of these children inflammatory activity continues into adult life.

This review discusses the problems facing adolescents with juvenile idiopathic arthritis (JIA) and how they are managed. Similar problems face adolescents with juvenile dermatomyositis, systemic lupus erythematosus, rarer connective tissue diseases and other musculoskeletal conditions, but a detailed discussion concerning these diseases is beyond the scope of this article.

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Key Points

- Approximately 20% of children with JIA will be unable to perform some aspect of self-care; the longer the follow-up, the greater both the disability and the deterioration in psychosocial function. Unemployment is common and, significantly, does not correlate with educational achievement. While many people with major disabilities enjoy happy, fulfilled lives, others with much milder functional impairment lead seriously restricted lives limited by factors, often simple, that may be avoided.

Now that 90% of children with chronic disabilities reach their 20th year, attention has been focused on their successful transition to adulthood. The key elements have been identified for an effective transition programme for adolescents with various chronic illnesses leaving the paediatric service. While the key elements also apply to adolescents with JIA, the details of the ideal transition programme for this group are far from established. Only 20% of the paediatric rheumatology departments in the UK provide a transition service and their standards are variable. To appreciate the late

consequences of JIA, paediatric rheumatologists should gain experience with adults whose arthritis began in childhood.

Problems facing adolescents with arthritis

The transition from childhood through the physical and emotional changes to adult life, from a supporting family to independence in the community, and from school into employment, faces all adolescents. Any disability brings its additional burdens of depression and difficulty in coping, poor body image, problems with challenging parental authority, and a tendency to reject treatment. The specific additional problems with which adolescents with arthritis have to contend are highlighted in Table 2.

Psychological problems experienced by adolescents with juvenile idiopathic arthritis

The hopes of adolescents with arthritis for sexual experience, marriage and children are no different from those of their able-bodied peers, but their expectations are poor, with anxiety centering on physical appearance and whether they could attract a partner or bring up children. Parental attitudes are important as they are often transmitted to the child. They commonly believe that their child will not marry but hope for it, so someone ‘can look after him/her when we have gone’. Their natural concern to overprotect the child is neutering and infantilising. Some parents insist on choosing their children’s clothes, and seem to ‘dress them down’ to draw attention away from them. Despite their misgivings, many adolescents will realise their hopes. The frequency of premarital sexual intercourse is two-

Table 1. Classification of juvenile idiopathic arthritis (JIA).

<table>
<thead>
<tr>
<th>Subtype</th>
<th>Characteristics</th>
<th>Outcome</th>
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<tbody>
<tr>
<td>Systemic onset</td>
<td>Onset usually less than 7 years Systemic features: fever, rash, serositis Polyarthritis (often symmetrical)</td>
<td>Systemic features may recur In 50%, disease resolves with little disability In 50%, there is progressive involvement of many joints Moderate to severe disability Rarely amyloidosis</td>
</tr>
<tr>
<td>Oligoarticular</td>
<td>Up to four joints affected Onset usually less than 5 years</td>
<td>Joint disease usually ‘burns out’ May have residual limb length discrepancy 20% develop iridocyclitis</td>
</tr>
<tr>
<td>Extending oligoarticular</td>
<td>Onset as oligoarticular but progressive involvement of additional joints</td>
<td>Follows more aggressive course</td>
</tr>
<tr>
<td>Polyarticular: rheumatoid factor-negative</td>
<td>Presents at any age More than four joints affected, small joints often present first</td>
<td>Eventual disability related to number of diseased joints and duration of disease before adequate treatment Hip disease occurs in 50% and is associated with poor outcome</td>
</tr>
<tr>
<td>rheumatoid factor-positive</td>
<td>Rare Symmetrical, small joints with large joints also affected Subcutaneous nodules</td>
<td>Erosive, destructive disease Moderate to severe disability</td>
</tr>
<tr>
<td>Enthesitis-related arthritis</td>
<td>Includes spondyloarthropathies, except psoriatic arthritis Axial and peripheral joints Rarely acute iritis High frequency of HLA B27</td>
<td>Often persists to adult life Very variable course Acute iritis may develop</td>
</tr>
<tr>
<td>Psoriatic arthritis</td>
<td>Oligo- or polyarticular arthritis before or succeeding onset of psoriasis Dactilitis and nail pits Family history common</td>
<td>Relapsing and remitting course Prognosis dependent on number of joints involved</td>
</tr>
</tbody>
</table>

In all these subgroups children most at risk for an unsatisfactory outcome are those referred late or who have had a long disease duration. Early recognition and management of complications are essential for optimal treatment and eventual quality of life. The use of methotrexate has revolutionised management and outcome for many of these subgroups.
thirds that of their healthy peers, the principal limiting factor being disease activity causing pain and fatigue rather than deformity.

Social isolation is common. One study\textsuperscript{11} showed that 50% of these children had no contact with any child outside their family during school holidays yet, significantly, the degree of isolation did not correlate with disease severity – many were fully mobile. A child may feel unattractive because of deformities, appearing younger than his/her age, neutered by lack of privacy in personal hygiene, and frequent, impersonal body contact by health professionals that creates a detached attitude towards intimacy.

The transition programme

A transition programme is an essential element in the care of these children and must effectively start with the parents on the first day of diagnosis, inculcating a ‘can do’ attitude and maintaining a realistically optimistic and positive approach throughout (Table 3). Approximately a year before the target date of transfer to the adult service the following concerns should be addressed:

- coping with the style of care provided by the adult service
- employment
- life away from the family home
- finding a partner.

A successful programme anticipates the possible obstacles and manages a smooth transition. Failure to do so can result in the patients being lost to follow-up, having a precipitant transfer to adult services by refusing to attend the paediatric clinic or losing school, or by other events such as pregnancy or suicide\textsuperscript{7}. Various models to manage the transitional programme have been tried, including an ‘adolescent physician’\textsuperscript{6,7} and a generic community-based transition service using the Young Disability Team\textsuperscript{6}. The favoured model in the UK is a transition programme coordinated between the paediatricians and adult physicians.

The transition programme is a process, not an event. Preferably, there should be a protocol in place for the programme so that no details are overlooked. The programme is necessarily multidisciplinary, so a single coordinator, possibly a nurse specialist, should be responsible and a check-list drawn up. Information can be disseminated by leaflets, paediatricians and adult rheumatologists, Young Arthritis Care, Arthritis and Rheumatism Campaign, The Lady Hoare Trust, out-of-hours clinics, parent support groups and social evenings.

Coping with the adult service

Paediatricians and patients may resist ending a respected and supportive, lifelong relationship by transferring to an adult service which may be viewed as ‘uninterested in paediatric conditions’.\textsuperscript{7} Coping with the adult service requires involving the adolescent in decision making and educating him/her about the disease, its treatment and disability. The emphasis is on taking responsibility for the management of the disease by:

<table>
<thead>
<tr>
<th>Key problem</th>
<th>Cause</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Impaired mobility/self-care</td>
<td>Any inflamed joint will affect mobility or an aspect of self-care in some way Osteoporosis</td>
<td>A major cause of residual disability in adolescents is the destroyed hip An increasing number of hip replacements are being performed in this group</td>
</tr>
<tr>
<td>Visual impairment</td>
<td>Steroid cataracts Chronic iridocyclitis</td>
<td>Iridocyclitis complicates 20% of oligoarticular disease ANAs are strongly associated If detected, the outcome is good All patients at risk must have regular ophthalmological reviews for the first 5 years of disease</td>
</tr>
<tr>
<td>Poor body image</td>
<td>Short stature Joint deformity Micrognathia Drug-induced obesity and hirsutism Delayed puberty</td>
<td>Disease activity, steroid medication and inadequate nutrition all contribute to short stature Exogenous growth hormone has been given with anecdotal success, but the literature is sparse</td>
</tr>
<tr>
<td>Low self-esteem</td>
<td>Poor body image Infantilisation and neutering by parents Social isolation</td>
<td></td>
</tr>
<tr>
<td>Unemployment</td>
<td>Impaired mobility Lack of education Poor motivation Attitude of employers</td>
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\textsuperscript{ANA} = antinuclear antibody.
encouraging self-medication

• adolescents holding drug monitoring forms

• sending copies of clinic letters to the patients themselves, and

• seeing the patients separately from their parents.

The final transfer must be sensitively managed. Information about the service, such as names, photographs and the site, in the form of leaflets or posters displayed in the paediatric waiting areas raises expectations and allays uninformed fears. The adolescent may be introduced to the adult rheumatologist well before the expected date of transfer, either in a joint clinic or by a visit to the adult service with a trusted member of the paediatric team.

Living independently

It is often difficult for a parent to ‘let go’ of a child who has been so dependent. It is well recognised that a degree of independence is paramount to a child’s development. Disability living allowance, housing allowance and transport need to be addressed when an adolescent plans to leave home.

References


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