PROBLEMS OF STILL'S DISEASE IN ADULT LIFE*

BY

U. STEIGER

Basel

Although Still's disease is relatively uncommon and the ultimate prognosis is not as bad as at one time thought (Edström, 1958; Ansell and Bywaters, 1959; Schlesinger, Forsyth, White, Smellie, and Stroud, 1961), nevertheless it can lead to a slow accumulation of crippled adults. During the past 6 years, eleven patients now adult who had suffered from Still's disease, together with five cases of Still's disease seen before the age of 16, and 289 cases of adult rheumatoid arthritis have been referred. This paper is based on the eleven patients seen late in the course of their disease.

Ten fulfilled the diagnostic criteria of definite Still's disease (Ansell and Bywaters, 1959) while the eleventh case initially fulfilling these criteria had developed ankylosing spondylitis (Case 9). The age at the time of referral varied from 17 to 49 years (Table). Activity of the disease was designated '0' if the erythrocyte sedimentation rate was below 10 mm./hr, '+' from 10 to 50 mm./hr, and '++' over 50 mm./hr. Despite a duration of 43 years, active disease may still be present although there is a tendency for this to diminish as time passes. Apart from active disease, the main reason for referral was involvement of the weight-bearing joints, especially the hip and knee joints; only two patients (Cases 3 and 6) who had generalized deformities had poor function in the shoulders, elbows, and hands.

The complication of recurrent iridocyclitis was present in three cases. Two patients had developed this soon after the onset of their disease and the third (Case 9) much later. For the first 20 years of the disease this patient had typical Still's disease with fever and severe arthritis; during this time he developed complete ankylosis of both hips and knees and all joints of the feet (Fig. 1). Not until the age of 26 did he show evidence of ankylosing spondylitis with the later involvement of the right shoulder and the development of an aortic valvular lesion.

![Figure 1](http://ard.bmj.com/)

**Fig. 1.—Case 9. Pelvis of man aged 36 years, 30 years after the onset of Still's disease, and 10 years after the first clinical signs of ankylosing spondylitis.**

---

*Read at a meeting of the Heberden Society at Taplow on June 17, 1967.

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs)</th>
<th>Duration (yrs)</th>
<th>Reason for Referral</th>
<th>Active Disease</th>
<th>Employment</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>At Referral</td>
</tr>
<tr>
<td>1</td>
<td>17</td>
<td>3</td>
<td>Taral joints</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>2</td>
<td>20</td>
<td>5</td>
<td>Hip</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>3</td>
<td>20</td>
<td>8</td>
<td>Severe deformities</td>
<td>++</td>
<td>0</td>
</tr>
<tr>
<td>4</td>
<td>17</td>
<td>9</td>
<td>Deformities</td>
<td>+</td>
<td>0</td>
</tr>
<tr>
<td>5</td>
<td>19</td>
<td>13</td>
<td>Right hip, neck</td>
<td>+</td>
<td>partly</td>
</tr>
<tr>
<td>6</td>
<td>20</td>
<td>18</td>
<td>Severe deformities</td>
<td>+</td>
<td>0</td>
</tr>
<tr>
<td>7</td>
<td>22</td>
<td>19</td>
<td>Both hips</td>
<td>0</td>
<td>+</td>
</tr>
<tr>
<td>8</td>
<td>25</td>
<td>20</td>
<td>Knees, recurrent iridocyclitis</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>9</td>
<td>36</td>
<td>30</td>
<td>Ankylosing spondylitis, aortic valvular lesion, iridocyclitis</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>10</td>
<td>48</td>
<td>32</td>
<td>Severe deformities</td>
<td>0</td>
<td>partly</td>
</tr>
<tr>
<td>11</td>
<td>49</td>
<td>43</td>
<td>Knees, iridocyclitis</td>
<td>+</td>
<td>+</td>
</tr>
</tbody>
</table>
Three histories are given in more detail.

**Case 6, a girl, now aged 21**, had an acute onset of typical Still's disease at the age of 2 years and 2 months. Up to the age of 12 she spent the greater part of her life in a children's hospital and finally attained a fairly good condition on corticosteroid therapy. In the following years her health deteriorated and she had no proper care until the age of 19 when she was admitted to an orthopaedic hospital. She was then unable to look after herself and could not walk; she had ankylosis of the left elbow and flexion and adduction contractures of the right hip joint. After an arthroplasty of the left elbow and a Girdlestone resection of the right femoral head, she is now able to walk and to take care of herself. At present she has mildly active disease that can be controlled with salicylate and she is following a rehabilitation programme.

The x rays of the hand shows complete fusion of the radius, carpal, and metacarpal bones but with a good joint space at the carpometacarpal joint of the thumb (Fig. 2). The hands are badly deformed but there is a satisfactory grip and use of the finger tips (Fig. 3). There was complete fusion of the left elbow (Fig. 4), but after arthroplasty good function was restored (Fig. 5).

---

**Figure 2.** Case 6. X-ray of hands (see text).

**Figure 4.** Case 6. Complete fusion of left elbow joint.

**Figure 3.** Case 6. Satisfactory grip and use of finger tips.

**Figure 5.** Case 6. Flexion and extension after arthroplasty of left elbow.
Case 10, a 53-year-old woman, had an acute onset of polyarthritis with fever at the age of 15. She was confined to bed for several prolonged periods with short intervals of partial mobilization. From the age of 32 onwards there has been no further evidence of active disease and in spite of severe deformities of the hands, hip, and knee joints she has been able to work regularly at home. For several years she has had increasing difficulty in walking because of severe flexion deformities of the hip joints and valgus deformity of the knee joints (Fig. 6a). The valgus deformity of the knees led to stress fractures of both fibulae (Fig. 7, opposite).

The patient underwent a programme of surgery lasting almost 2 years with a Smith-Patersen cup for the right hip joint, osteotomy of the left femoral condyle, and Platt arthroplasty of the right knee. Her posture (Fig. 6b) and ability to walk have very much improved. Despite severe deformities of the hand with complete ankylosis of all the interphalangeal joints (Fig. 8), satisfactory function is preserved; her part-time work consists of assembling watches.
and effusion in the knees. Since then he has had recurrent trouble, predominantly in the knees, but is in full-time employment as a laboratory technician.

Radiologically, there is an increase in size of the radial head due to a growth disturbance (Fig. 9). A needle biopsy of the knee joint gave synovial membrane showing an active process 44 years after the onset of the disease and 22 years after the onset of recurrent effusions. Radiologically there is secondary osteo-arthrosis but satisfactory alignment has been maintained. Treatment consists of preventing flexion deformity of the knees, regular quadriceps exercises, and occasional local corticosteroid infiltration.

Summary

These patients can be divided into two groups. Cases 1 to 8 ranged from 17 to 25 years of age at the time of referral and Cases 9 to 11 from 36 to 49 years of age. The main problems in the younger

Fig. 7.—Case 10. Severe valgus deformity of knee joint caused by collapse of tibial plateau. Note stress fracture of fibula.

Case 11, a man now aged 50, had an acute onset of typical Still's disease at the age of 6; soon afterwards he developed severe iritis in the right eye culminating in unilateral blindness. He was treated in hospital for one year and about 2 years later his condition improved. He had no significant joint trouble until the age of 26 when he had a recurrence of pain with soft tissue swelling

Fig. 9.—Case 11. Growth deformity of head of radius.

Fig. 10.—Case 6. Crippling deformity, i.e. flexion contracture of hip and knee joints and severely ankylosed pes equinus.
age group were active disease and difficulty in adapting to deformities. During the time these patients were under my care, Case 3 had hand surgery in addition to general treatment and both this patient and Case 4 became able to do part-time work. Patients in the older age group were referred on account of increasing deformity in damaged joints. Deformity rather than active disease was the main reason for incapacity for work.

The importance of the prevention or early correction of deformities in Still's disease has been emphasized by Ansell (1965, 1966). In most cases contracture of the knees and poor position of the wrists can be avoided by proper splinting and exercise programmes, as can pes equinus (Case 6, Fig. 10, p. 149); complete ankylosis of the elbows is also preventable by regular physiotherapy. Late correction of deformities being often difficult or impossible, every effort to prevent crippling deformities must be made throughout the patient's illness.

My thanks are due to Dr. B. M. Ansell for her help and advice in writing this paper.

REFERENCES


Le problème de la maladie de Still pendant la vie adulte

RéSUMÉ

On peut diviser ces patients en deux groupes. L'âge des Cas 1 à 8 se rangeait entre 17 et 25 ans et celui des Cas 9 à 11 entre 36 et 49 ans au moment de la première entrevue. Dans le groupe plus jeune l'activité morbide et l'adaptation difficile aux difformités constituaient le problème principal. Pendant le temps que j'avais le soin de ces malades, le Cas 3 a été soumis à la chirurgie de la main tout en recevant son traitement général et ce malade, ainsi que le Cas 4 sont devenus capables d'un travail partiel. Les patients du groupe plus âgé ont été commis à nos soins en raison de la difformité plus accentuée de leurs articulations. C'était cette difformité plutôt que l'activité morbide qui les empéchait de travailler.

L'importance de la prévention ou de la correction rapide des difformités dans la maladie de Still a été soulignée par Ansell (1965, 1966). Dans la plupart des cas la contracture du genou et la difformité du poignet peuvent être évitées par des attelles appropriées et par des programmes d'exercice; il en est de même pour le pied bot équin (Cas 6, Fig. 10); on peut aussi prévenir l'ankylose complète du coude par une physiothérapie régulière. Sachant qu'une correction tardive des difformités est souvent difficile ou impossible, il faut s'efforcer de les prévenir pendant toute la durée de la maladie.

El problema de la enfermedad de Still en la vida adulta

SUMARIO

Estos enfermos pueden repartirse en dos grupos. La edad de los Casos 1 a 8 se extendía de 17 a 25 años y la de los Casos 9 a 11 de 36 a 49 años al tiempo de la primera entrevista. En el grupo más joven la actividad morbosa y la dificultad de adaptación a la deformidad constituieron el problema principal. Durante nuestro cuidado, el Caso 3 fue sometido a la cirugía de la mano además del tratamiento general y este enfermo, así como el Caso 4, se volvieron capaces de trabajar parcialmente. Los pacientes del grupo más maduro nos fueron enviados a causa de deformidad creciente de las articulaciones afectadas. La deformidad más bien que la actividad morbosa fue la causa principal de la incapacidad de trabajo.

La importancia de la prevención o de la corrección de las deformidades en la enfermedad de Still fue subrayada por Ansell (1965, 1966). En la mayoría de los casos la contractura de la rodilla y la deformidad de la muñeca pueden evitarse con el uso de aparato ortopédico y de un programa de ejercicios; lo mismo se aplica al pes equinus (Caso 6, Fig. 10); se pude también evitar la anquilosis completa del codo por fisioterapia regular. Sabiendo que una corrección tardía de deformidades es a menudo difícil o imposible, se deben hacer esfuerzos para prevenirlas en todas las fases de la enfermedad.