Spondylitis Ankylopoietica

Synonyms: Strümpell-Marie Disease, Spondylosis Rhyzomelique, Spondylitis Deformans, Rheumatoid Spondylitis, Poker Spine, Spondylitis Adolescence, etc.

This condition has received considerable attention by various investigators during the past few years, and much has been added to the scientific knowledge of an affliction that has for long been recognized. It is surprising that, despite the long recognition of this disease, so little progress has been made towards determination of the true pathology, recognition with greater accuracy of the clinical picture, and particularly towards interpretation of the roentgenographs and the significance of roentgen therapy.

The slowness of the pathological investigation is readily understood when one considers the fact that few patients ever come to necropsy in the early or even moderately advanced stages of the disease. The disease in itself is not of a fatal character, and sufferers may reach an advanced age, when the process has "burned itself out", and only the permanently deformed wreck has been left.

Clinically these cases run to quite a uniform pattern, and although the onset and development may vary as to age and character, the onset in the majority occurs during adolescence, 17 to 18 years, with characteristic complaints of a stiff, sore back, without significant concomitant injury. The disability is steadily progressive. Coincidentally the patient may complain of pains and aches in some of the joints, particularly the shoulders and hips. The condition occurs predominantly in males and frequently in men who have been of the athletic type, indulging in baseball, hockey, swimming, etc. Such activities at first have to be curtailed and in the course of a few years entirely abandoned. Associated with this disability
there is usually a tired, rundown feeling and postural discomfort. Any jarring causes extreme discomfort in the back. Pain radiating along the intercostal nerves is also a common symptom. Some attribute the onset to a minor back injury, but this is not characteristic.

Physical examination in the main elicits three findings: (1) under-nourished, sometimes almost emaciated appearance; (2) limitation of movements of the spine; and (3) moderate to marked diminution of chest expansion. (The blood sedimentation rate is variably increased. Iritis also occurs in this disease.)

Figures 1, 2, 3 and 4 demonstrate the appearance of such a case. This man, J.H.M., age 27, shows the slightly stooped posture in figure 1. Figure 2 demonstrates the slight list to the left, and figure 3 demonstrates the limitation of movement of the spine as he attempts to overcome the list. Figure 4 indicates the limitation of flexion of the spine, all movement taking place at the hip joints. The back has then truly become a poker-like rod. However, this rigidity as a feature of the condition may make its appearance early, and judging from the effects of bed-rest, it is then due mainly to muscle spasm. Chest expansion at the first examination on this individual was no more than one-quarter of an inch, and may be put down as nil, since so many patients have a tendency to throw their shoulders back at the limit of expansion, thus giving the false impression of at least some mobility of the thoracic cage, or of exaggerating the true natural mobility. The undernourished appearance of this individual is amply demonstrated.

Blood sedimentation rates have been determined by the Cutler method and the index propounded by Dr. J.D. Adamson has been used, the normal being -12 to +12.

Roentgenography - Three features of this aspect obtain in diagnosing these cases: (1) the well advanced case is demonstrated in figures 5
and 6. These show calcifications or ossification of the anterior and lateral spinal ligaments in the lumbar region. (I might explain here that although photographs were made from these negatives, they do not demonstrate nearly so well the characteristic lesions). (2) Figure 7 demonstrates the already moderately advanced lesion in the sacro-iliac joints. At the beginning this consists of a fuzzy appearance of the articulating edges with a zone of rarefaction, which changes progressively to a zone of sclerosis and then to partial or complete fusion, as in figure 8. Figure 9 shows still more clearly the complete fusion of the sacro-iliac joints. The significance of this characteristic has been emphasized by Blair (1) and Smyth and Freyburg (2) and will be alluded to again. (3) The third feature, that of apophyseal joint involvement, is much more difficult to interpret and demonstrate. Oppenheimer (3) has only recently drawn attention to this. Its detection compels taking oblique views in both directions, and necessarily of both the lumbar and thoracic spine, to demonstrate the extent of involvement, if any, of these articulations. The nature of the variable angle of obliquity of these articulations in different individuals sometimes calls for repetition of films, which is not only time-consuming but costly as well.

**Diagnosis:** Many people complain of various types of backache, but I know of none that follows such a characteristic and uniform pattern as does Spondylitis Ankylopoietica. True, in the early stages nothing may be demonstrated except stiffness and aching of the back, but this is usually of a progressive character. Most commonly the condition has its inception in adolescence and I am in complete accord with Smyth and Freyburg, quote: "It has been emphasized that sacro-iliac arthritis often (but not always) is the earliest roentgen sign of this disease. In all our patients with early disease with roentgenographic abnormalities present only in the
sacro-iliac joints, the existence of marked spinal motion restriction, diminished chest expansion, progressive loss of weight and disability, with increased blood sedimentation rate - made the diagnosis." In the more advanced stages the spine becomes so rigid that there is no doubt about the diagnosis. The degree of deformity varies tremendously and is probably influenced by the position which these patients assume to get relief. A sagging bed is an important factor here.

Concisely, then, if a patient presents himself complaining of a progressively painful and stiff back, rundown condition, with loss of weight, and there is roentgenographic evidence of sacro-iliac joint changes, and diminished chest expansion, a diagnosis of Spondylitis Ankylopoietica is almost certainly established, and correctly so in the vast majority of instances.

From a diagnostic standpoint one can infer that recognition of the characteristics of this condition has led to more frequent diagnosis. There is, according to our index, a veritable paucity of these cases in the years preceding 1939, and a study of some of the histories reveals that the diagnosis was based on roentgenographs of only the lumbar or thoracic spines or both. Certainly no mention is made of the sacro-iliac joints as a diagnostic feature. The cases, numbering fifteen, comprising the material for this paper have been diagnosed between the years 1939 and 1944 inclusive, and the apparent increase, I am sure, can only be attributed to our improved methods of diagnosis. Even so, the investigation of these cases has been incomplete and in future a more thorough one will be conducted, although from the diagnostic standpoint that which has been done appears to have been sufficient.

Etiology: At present there is speculation regarding the actual cause of this condition. Its association with conditions as gonorrhoea or other forms of infection, polyarthritis, rheumatoid arthritis, trauma, etc.,
is of course no explanation. Likewise the common incidence during the
age of adolescence gives us no more information.

<table>
<thead>
<tr>
<th>TABLE I</th>
</tr>
</thead>
<tbody>
<tr>
<td>No associated disease</td>
</tr>
<tr>
<td>No. of Cases</td>
</tr>
<tr>
<td>Total</td>
</tr>
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</table>

#D.K.

Table I indicates the relative frequency of rheumatoid arthritis either as a forerunner or a concomittant factor. To establish the cause of rheumatoid arthritis will probably suffice for both. The existence of a focus of infection, specific or otherwise, would indicate that this cause is either toxic, or infective, or a combination of both. The presence of myositis ossificans, I believe, points to a metabolic disturbance of a physio-chemical imbalance as a probable factor.

Blair has significantly termed both Spondylitis Ankylopoietica and rheumatoid arthritis as being sulfur demineralization processes. His explanation that the loss of resiliency of the intervertebral discs is due to organic sulfur depletion is at first logical. However, the only proof of such demineralization is an experimental one based on the response of mast cells to roentgen therapy.

It is found that all forms of fibrous tissue, such as ligaments, tendons, etc., are rich in mast cells. The staining property of normal

#One case has been added to this series because it demonstrates the associated condition of myositis ossificans.
mast cells demonstrates the presence of metachromatic granules. Following roentgen therapy these metachromatic granules disappear, and on their disappearance is based the theory of liberation of sulfur. Experimentation has also shown that mast cells are secretory in function and liberate heparin, which is an anticoagulant, and that this is composed in part at least of sulfur.

The clinical application of this discovery is that mast cells give up the sulfur contained within them and the sulfur is transported to sulfur depleted structures with re-establishment of resiliency. The natural deduction from this is to the effect that the osseous fixation of the spine is confined to the fibrous and fibro-cartilaginous structures about the spine.

This could be more readily accepted if in each and every case of spondylitis Ankylopoetica ligamentous calcification or ossification could be demonstrated, and particularly if it could be shown to be in direct ratio to the degree of spinal immobilization. However, Blair himself stresses the early involvement of the sacro-iliac joints and its early diagnostic significance.

Oppenheimer refutes both these theories by demonstrating that the earliest lesion is in the apophyseal articulations and that sacro-iliac joint changes as well as ligamentous changes may be absent. He attributes the whole process to arthritis involving the apophyseal articulations and believes that the other features are secondary. Furthermore, it is at the pre-sacro-iliac and pre-ligamentous involvement stage that roentgen therapy has its greatest application. On the other hand, Smyth and Freyburg even suggest that the increased ligamentous calcification may not result from continued active inflammation but may actually be a part of a healing process.

It would appear, therefore, that a variety of causes may initiate this condition: toxicity, infectivity, metabolic and physico-chemical disturbances and even trauma. What part sulfur plays in the process may be only
a matter of cause and effect, yet its depletion, resulting in lost resiliency, may still be a vital factor. Certainly I believe there is no distinction between the nature of rheumatoid arthritis and Spondylitis Ankylopoietica except by their manifestations. Why the former does not respond to roentgen therapy and the latter does is a mystery to me.

The order of involvement of the various articulations, namely sacro-iliac, lumbar, cervical and thoracic, in that order, is in direct relationship to the amount of strain to which these joints are normally subjected. This is the significance of the roentgenographic evidence of sacro-iliac joint changes as the first sign. Variations in this order may at least in part be due to the fact that the amount of strain varies in different individuals.

**Incidence:** There seems no way in which to determine with any degree of accuracy the incidence of Spondylitis Ankylopoietica in the general population. Nor, perhaps, does it matter, but I have been impressed by the number of cases diagnosed in this Clinic during the past five years, namely fifteen, as compared with the number of cases of Pott's Disease of the spine, over the same period: two definite cases and one doubtful. The incidence of Spondylitis Ankylopoietica is 0.5% and for Pott's Disease of the spine 0.1%. To keep these figures as unprejudiced as possible only new cases presenting themselves at this Clinic over the last five-year period have been considered. Any cases previously diagnosed but returning for treatment for that or some other condition, have not been included.

That this disproportion actually exists I am not prepared to admit. In my mind even a suggestion of such relative frequency is of the utmost importance, since Spondylitis Ankylopoietica has heretofore been given comparatively little recognition, to the point where the general practitioner usually knows it only by the term Poker Spine. The average medical student
has even less knowledge of it.

This matter should be rectified and more stress laid on it by the clinician. The importance of doing so is amply expressed by S. Gilbert Scott, England, Radiologist, describing this type of patient, who, if taken in time, can be saved, but if neglected either through misdiagnosis or ignorance or delay, will become what he calls a "Spondylitic Wreck".

The argument has been advanced that many cases of Spondylitis Ankylopoietica which have existed for years may only now be diagnosed, implying an exaggerated incidence. This I do not completely deny, but out of this series of fifteen cases six dated the duration of their symptoms as four years or less, so that although the incidence may not be on the increase, these patients do appear to be presenting themselves for diagnosis at an earlier period. The chronicity of the nature of Spondylitis Ankylopoietica naturally permits that these cases may be diagnosed only years after the onset, but to some extent this argument also applies to Pott's Disease of the spine, for in the occasional case one finds that nature has in part bridged the diseased vertebrae by new bone, permitting sufficient immobilization and partial arrest of the disease, thus postponing the patient's submission to examination or treatment.

Treatment: Previously the treatment in these cases was in the main palliative, although braces and casts as a corrective measure have been used. The braces are usually inadequate, and corrective casts, besides being tedious and cumbersome, are difficult to endure. One case record in our files reveals rather extensive surgery as a form of treatment, which was apparently successful. The laminae were completely
fused in the lumbar region, and after creating an interval between them by excision of bone it was possible to mobilize the spine so that the interval of five-eighths of an inch between the spinous processes previous to operation could be reduced to one-quarter of an inch. This was followed by a series of corrective casts and resulted in improved posture.

It is the consensus of opinion that once the spine becomes a rigid rod, any associated pain subsides. Though this may be true in theory, it is not universally borne out in practice, as some patients continue to complain of pain in various regions, mainly along the intercostal nerves, and of postural discomfort.

Roentgen therapy has now been fairly generally accepted, and not without reasons. Whatever the rational of such therapy, the results have been very encouraging; but several courses may be necessary either to obtain the maximum results or because of degrees of recurrence or continued progress of the condition. This, however, is only part of the treatment in a well managed case, and supplementary orthopaedic measures constitute the balance of the treatment. There seems to be no set dosage of roentgen therapy, and the radiologist is apparently of the opinion that, in order to get the desired result, concentration with a cumulative effect over the affected area is most desirable. That this may be logical is suggested by Comroe (5), who believes that the marked dilatation of the capillaries, subcapillary plexuses and arterioles under the area treated may be part of the answer to the improvement from roentgen therapy. Blair treats the whole body with low voltage and wide dispersal on the basis of liberating sulfur or heparin from the mast cells, thus replenishing the depleted sulfur from the affected tissues.
If capillary dilation and hyperaemia is the basis of roentgen therapy then concentration over the affected area seems the logical procedure.

Baker uses bed rest in conjunction with roentgen therapy, employing a reversed Gatch frame so that the break intended for under the knees is placed under the thoracic kyphos. This achieves two objects, muscular relaxation and corrective posture.

Swain has successfully used body jackets with increasing degrees of correction. The few cases which I have had the privilege of treating have been done by a combination of all these measures. Roentgen therapy has been administered with consideration of both sulfur and hyperaemia as the underlying factors, and the torso only has been subjected, giving a total of 1,100 roentgen units, 100 units daily for 11 days. This gives approximately 1,000 units, as there is always some wastage.

The Gatch frame has been used where the patient was hospitalized, and if at home a pillow placed under the thoracic kyphos and boards under the mattress. In those cases where sufficient mobility was present and it seemed that exercises would be beneficial, they were instituted. The plan of the apparatus demonstrated by Baker was used and an accompanying diagram is presented. The mechanics
of this device consist in fixation of the pelvis with the strap placed at or near the level of the kyphos and acting as a fulcrum; by throwing the part of the body above the kyphos backwards some degree of mobility is re-established.

In one case where there was so much rigidity of the spine that exercises appeared not to accomplish anything, a corrective jacket was applied, and he is at present wearing it. At the end of one month's time another jacket with still further correction will be applied, and this repeated until what appears the maximum correction has been obtained.

The purpose in using the cases presenting themselves over a five year period is an arbitrary one, for few cases diagnosed as Spondylitis Ankylopoietica are listed in the previous years. The associated myositis ossificans as shown in Table I was in a case, D.K., and actually not one of this series, who sustained a dislocation of his left hip in 1930 which necessitated open reduction and which was followed by terrific myositis ossificans and ultimately led to repeated amputations at increasing higher levels of that extremity. It was only accidentally noticed in 1943 upon taking a plate of his pelvis that his sacro-iliac joints were fused, and upon this assumption the lumbar and thoracic spine were roentgenographed, showing typical Spondylitis Ankylopoietica.

Our index reveals that over the years 1917 to 1938 inclusive a diagnosis of Spondylitis Ankylopoietica was made on only six cases, inferring that our knowledge of this condition has increased considerably along with our methods of diagnosis.

There are two incidental features of this investigation which have impressed me. One is the effect of roentgen therapy on the blood sedimentation rate. Whereas the sedimentation rate was increased prior to roentgen therapy, immediately following the therapy the sedimentation
rate was increased still further. Discussion of this fact with one of the Roentgenologists revealed that he had no knowledge of this effect. The blood sedimentation rate is governed by the blood globulin content. Since globulin is manufactured in the liver, roentgen therapy may lead to stimulation of globulin production by virtue of liver stimulation and thus account for the increased rate. The second feature concerns the sulfur content estimation in the human body. Biochemistry manuals state that the blood sulfur molecule is so complex and the content so variable that no standard can be arrived at. It has been suggested to me that if some of the tissues which are likely to have a fairly stable sulfur content were analyzed, such as the nails and hairs, a normal standard could probably be arrived at. The sulfur content of these tissues in an affected patient could then be determined and might be a step towards proving or disproving the theory of sulfur demineralization as the underlying factor in Spondylitis Ankylopoietica.

**Conclusion:**

<table>
<thead>
<tr>
<th>Patient</th>
<th>Sex</th>
<th>Age</th>
<th>Duration of symptoms in years</th>
<th>Age of onset</th>
<th>Blood sedimentation rate</th>
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<tbody>
<tr>
<td>P.G.</td>
<td>M</td>
<td>37</td>
<td>19</td>
<td>18</td>
<td>18</td>
</tr>
<tr>
<td>E.M.</td>
<td>M</td>
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<td>9</td>
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<tr>
<td>S.U.</td>
<td>M</td>
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<td>2</td>
<td>25</td>
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</tr>
<tr>
<td>J.T.</td>
<td>M</td>
<td>22</td>
<td>3</td>
<td>19</td>
<td>19</td>
</tr>
<tr>
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<td>M</td>
<td>31</td>
<td>16</td>
<td>15</td>
<td>15</td>
</tr>
<tr>
<td>P.B.</td>
<td>M</td>
<td>49</td>
<td>29</td>
<td>18-20</td>
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</tr>
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<td>M.R.</td>
<td>M</td>
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<td>1</td>
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<tr>
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<td>M</td>
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<td>12</td>
<td>12</td>
<td>12</td>
</tr>
<tr>
<td>F.D.</td>
<td>M</td>
<td>38</td>
<td>3</td>
<td>35</td>
<td>35</td>
</tr>
<tr>
<td>T.S.</td>
<td>M</td>
<td>24</td>
<td>4</td>
<td>20</td>
<td>20</td>
</tr>
<tr>
<td>M.F.</td>
<td>M</td>
<td>27</td>
<td>15</td>
<td>12</td>
<td>12</td>
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<tr>
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<td>+38</td>
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<tr>
<td>J.L.D.</td>
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<td>17</td>
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<td>+35</td>
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<tr>
<td>H.J.M.</td>
<td>M</td>
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<td>3</td>
<td>24</td>
<td>+21</td>
</tr>
<tr>
<td>J.H.</td>
<td>M</td>
<td>31</td>
<td>8</td>
<td>23</td>
<td>+35</td>
</tr>
</tbody>
</table>
All the cases were males.

Average age at which the patient presented himself at this Clinic was 32.2 yrs.

Average duration of symptoms was 9.9 years.

Average age of onset was 22.2 years.

Blood sedimentation rate was ascertained in four cases and averaged 43.2.

One or two interesting sidelights respecting these cases might be mentioned. Case T.S. is one in which the only positive clinical, physical or roentgenographic findings consisted of sacro-iliac joint involvement. Sacro-iliac tests were all positive and the roentgenographs showed bilateral involvement of these joints progressing towards fusion as demonstrated by a six weeks' time interval. The spine was in all respects negative, including the apophyseal articulations. This patient was treated by surgical fusion of these joints and two months post-operatively is relieved of all symptoms. My opinion is that this is Spondylitis Ankylopoietica which has remained localized, or if it is progressing elsewhere it is doing so at a very slow rate. A blood sedimentation rate on him was +6 and would argue against any systemic reaction and substantiate the localization of the process.

F.D. was referred, having been diagnosed osteoarthritis of the spine. The fault I found in this was the fact that the referring doctor actually considered these two conditions to be one and the same. An opportunity afforded itself to explain the difference, and it was also learned that the patient was feeling far better following the roentgen and orthopaedic therapy than he had for years.

P.G. and J.M. were pitiful cases due to the terrific deformities, particularly in the latter. He had good use of his upper limbs, but his back was as rigid as a poker from the epistropheus down and his hip joints almost completely ankylosed. He was completely bed-ridden. An attempt at restoration of hip joint movement was very discouraging due to the atrophy
of the bones. A Vitallium Cup arthroplasty was done on the right hip and
a reconstruction operation on the left. Roentgen therapy was not attempted.

J.T. had a focus of infection, namely in the mandible following
an abscessed tooth, which was so intimately associated with the onset of
his back trouble that one cannot deny some relationship. The history is
that in July 1939 he had a left lower molar extracted, which began suppura-
tating about a week later, and though the gum was incised he obtained no
relief. Subsequently the mandible was drained externally and continued to
drain until May 1940. At this time the sinus closed, and he dates his
back trouble to the time of its closure. It appears to be only a coinci-
dence that the back trouble manifested itself the same length of time after
the formation of the focus of infection.

S.U. has been followed for a period of seventeen months with
roentgen therapy and corrective plaster jackets. Despite this therapy the
sacro-iliac joint condition has progressed until fusion is almost complete.

Three of these cases demonstrate one or both hip joint involvement.

There are probably some uncertainties regarding the prognostic
features associated with Spondylitis Ankylopoietica. There is certainly
a great variation in the degree of deformity or extent of involvement
resulting from this condition. It may be that where sacro-iliac joint
manifestation is the only demonstrable feature, the condition may remain
localized. Other more extensive manifestations may similarly remain local-
lized, or the process may become spontaneously arrested. Periods of remis-
sion apparently occur. A variation in disability and degree of deformity
may be due to this or the arrest of the process, and hence many cases may
never even be recognized.
Summary: This paper discusses the clinical, physical and roentgenographic aspects of the disease, setting forth definite diagnostic features.

The etiology is discussed from the standpoint of sulfur demineralization and arthritis.

Associated conditions, including rheumatoid arthritis, focus of infection and myositis ossificans, are mentioned.

A comparison between the incidence of it and Pott's Disease of the spine is attempted.

Fifteen cases comprise this series, of which only four have submitted to treatment.

Treatment includes roentgen therapy and various orthopaedic measures.

References

1. Blair, Harry C., M.D., Portland, Oregon, Spondylitis Adolescens - Strümpell Marie Disease, p. 663, S.G.O., March 1942
5. Comroe, B.I., Arthritis and Allied Conditions, Lea and Febiger, 1940