

ANKYLOSING SPONDYLITIS

A REVIEW OF 184 CASES

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WITH AN APPENDIX BY

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Some 5 years ago (Hart, Robinson, Allchin and MacLagan, 1949) we published a study of 73 patients with ankylosing spondylitis. In a follow-up study of these patients I have reviewed an additional 111 cases, personally examined, since that time. A study of these 184 patients (18 female, 166 male) forms the basis of this communication.

Symptomatology.—In this series of 184 cases the initial symptom was as follows:

<i>Symptom</i>	<i>No. of Cases</i>
Low backache	70
Low back stiffness without pain	18
Low back stiffness with slight pain	10
Buttock pains	21
Pain in "hips, groins or thigh"	16
Sciatica	9
Pain and/or swelling of knees	7
Pain and/or swelling of ankles, heels or feet	13
"Rheumatic fever"	4
Pyrexia of unknown origin with generalized aches and pains	2

Dorsal spine pain	6
Pains in shoulders	3
Head going forwards	2
Pains around the chest	3

Taking the first five headings together, 135 patients (73.4 per cent.) experienced pain and/or stiffness in the lower back and buttocks as the initial symptom as compared with 24 (13 per cent.) in whom the initial symptom lay in the periphery. The words "rheumatic fever" appear somewhere in the case history in eight instances in which it is clear that the disease was in fact ankylosing spondylitis.

In reviewing these case histories in detail it became clear that certain "small symptoms" were frequently present in the few months or years preceding the onset of the classical backache. If one accepted these early symptoms as part of the disease process, the age at onset ranged from 10 to 51 yrs in the male patients, and from 13 to 32 yrs in the female patients (Fig. 1). The discovery of sixteen

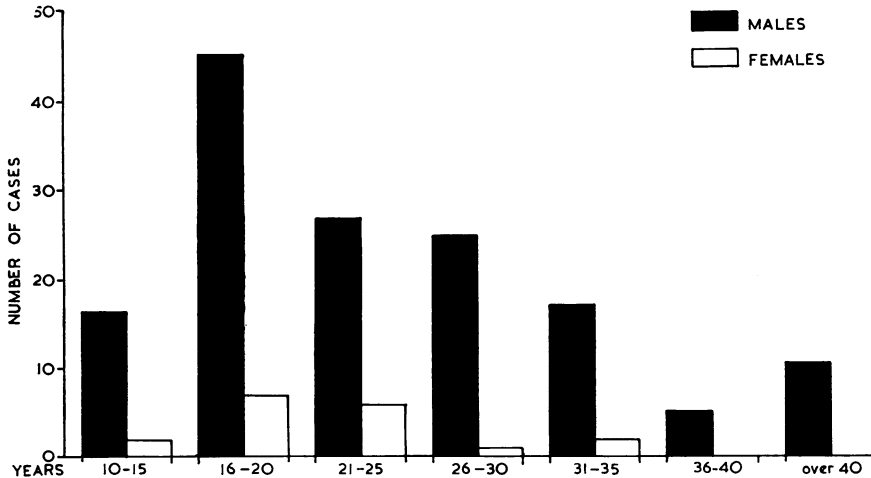


Fig. 1.—Age at onset of disease, by sex.

male patients in whom the early symptoms appeared between the ages of 10 and 15 years came as a surprise. It will be noted that 61 of these 184 patients developed the first symptoms of disease before their 21st birthday. Not infrequently complaints of intermittent "fibrositis" or occasional swelling of a peripheral joint marked the true onset of the disease. Such minor episodes appeared in the histories given by the patients at their first attendance and were not elicited by specific questioning; they appeared to be a part of this insidious relapsing-remitting disease rather than isolated and unconnected events. Three patients had made no complaint whatsoever and their spinal stiffness was discovered on routine examination. On closer questioning the typical history was forthcoming but the symptoms had been so slight that they had not been considered worthy of medical attention.

In four cases the initial pain assumed an erratic and peripheral course so that rheumatic fever was provisionally diagnosed. In two cases the acute, florid, extremely painful, clinical picture was seen, the patient being in too much pain to be out of bed but suffering extreme anguish at rest. The great majority presented in the usual manner, episodes of intermittent stiffness and aching in the back and buttocks gradually merging into the established disease pattern. Several secondary complaints were made later in the course of the disease. It is not intended to discuss these at length except to emphasize the frequency of bony tenderness, usually in the ischial tuberosities, but also in the brim of the pelvis, anterior superior iliac spines, greater trochanters, and occasionally sternum and ribs. This

ischial tenderness may be a major complaint, usually lasting only a few weeks or months or occasionally much longer. One of these patients had carried a small protective cushion with him for the last 2 years.

One point of interest that has appeared worthy of note is the occurrence in the spondylitic of short quite severe painful episodes, often lasting only a few days or one or two weeks. Such episodes are usually self-terminating and the sufferer will refuse admission to a hospital bed, saying he will be better in a few days. No particular precipitating cause is evident, but pain and stiffness may exacerbate to a marked degree either in one segment or, less frequently, throughout the entire spine. Such short episodes are in no way comparable with the exacerbation of disease seen in rheumatoid arthritis. Trauma does not appear to be the cause.

Physical Signs.—The classical physical signs are well known and do not need to be mentioned here. In 43 of these 184 patients peripheral joint swelling occurred at some time in the course of the disease in the following order of frequency:

Site	No. of Cases
Knees ..	28
Ankles ..	21
Feet ..	11
Wrists ..	11
Fingers ..	8
Elbows ..	4

Measurement of Spinal Movement.—This can only properly be done by using an instrument which eliminates hip and leg movement. The usual mode of measuring spinal flexion by measuring the dis-

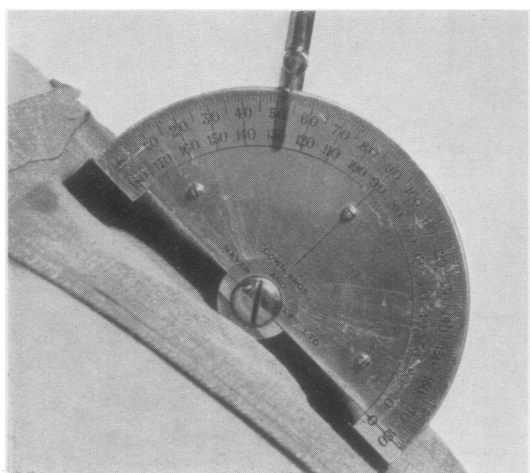


Fig. 2.—The Spondylometer.

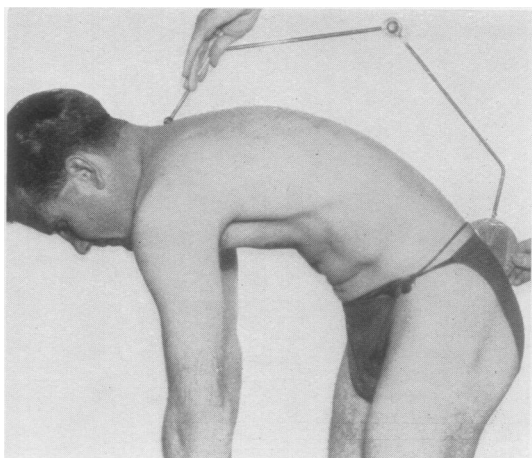


Fig. 3.—Measurement of spinal movement using the Spondylometer.

tance of the fingers from the floor with the spine in full flexion is notoriously unreliable as it measures not only spinal flexion but also hip mobility, pain and tenderness in hamstrings and knee and several other factors. For the past 4 years we have, therefore, been using a "Spondylometer", designed by Dr. W. F. Dunham (Figs 2 and 3; Dunham, 1949).

This instrument measures spinal movement between two points, the points we use being sacrum and vertebra prominens. Dunham has measured spinal movement in a number of normals and compared them with patients suffering from ankylosing spondylitis (Dunham, 1949). We have measured spinal range of movement in forty normal medical students, aged from 20-29 yrs, 5 ft. 5 in. to 6 ft. 3 in. tall. The spinal range lay between 75 and 120° and bore no relation to ability or inability to touch the floor with the fingers. No close parallel was observed between height and spinal range. These measurements are remarkably constant and provide the best measure of the progress of spinal stiffness in any given case. In early cases, where pain rather than structural change prevents movement, a definite increase can be obtained in the spinal range as a result of treatment by any effective method, whether it be cortisone, ACTH, or deep x-ray therapy, the response being much more rapid with the first two but less lasting (Hart, 1952). In most cases, however, there is a considerable degree of fixation of the spine and, though the pain may be greatly relieved by different forms of treatment, the spinal range of movement increases little, if at all. Fig. 4 illustrates spinal movement measured by the spondylometer in 143 patients at their first attendance at the clinic.

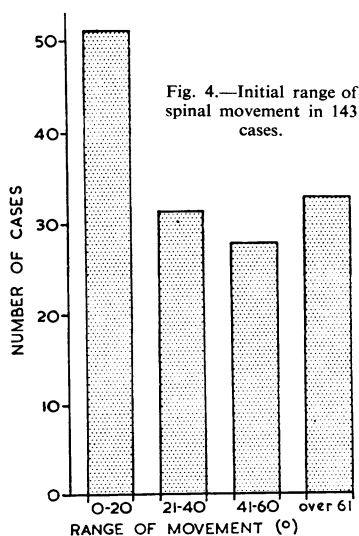


Fig. 4.—Initial range of spinal movement in 143 cases.

For the past 9 years we have, through the kindness of Dr. P. Hansell and the Photographic Department of the Westminster Hospital, taken clinical photographs of these patients (Fig. 5). Triple exposures illustrate certain points and serial photographs over the years enable one to record major changes in posture, but the method is not fine enough to use as a record of improvement or deterioration in posture or spinal mobility.

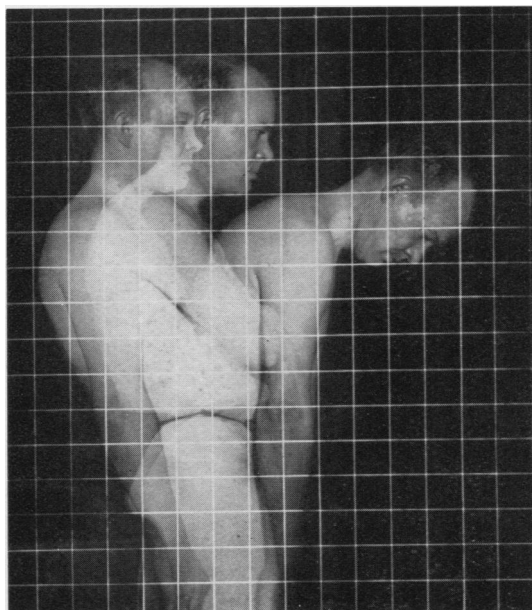


Fig. 5.—Triple exposure, showing range of spinal movement.

Thoracic Involvement.—A characteristic feature of ankylosing spondylitis is the reduced rib excursion which occurs as a result of fusion of rib with transverse process and body of vertebra. This reduction in intercostal respiration leads to over-use of the diaphragm and a double exposure skiagram of the chest in inspiration and expiration will, in such cases, demonstrate little rib movement but a generous diaphragmatic excursion (Hart, Bogdanovitch, and Nichol, 1950; Hart, 1950). The picture, therefore, differs from that seen in emphysema where both rib and diaphragm move little. This thoracic involvement is common in ankylosing spondylitis and though more frequent in advanced cases may occur as an early or even occasionally as a presenting physical sign. The resultant symptoms are usually stiffness of the chest wall, difficulty in fully expanding the chest, and an aching and discomfort on over-breathing, coughing, or sneezing. Tenderness and pain may also be felt in the sterno-manubrial region.

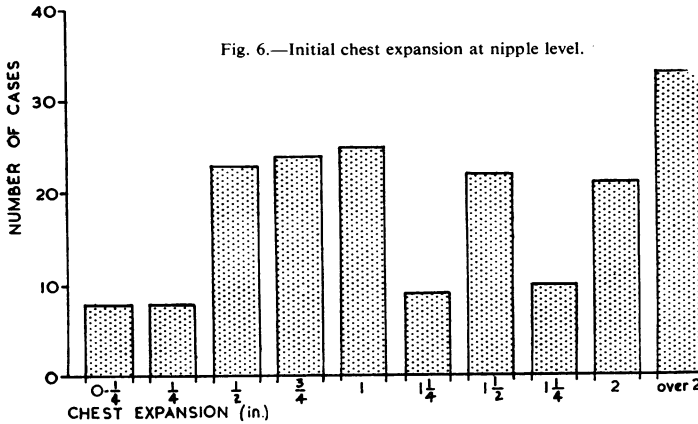


Fig. 6.—Initial chest expansion at nipple level.

In our series the initial chest expansion at nipple level, when first seen, was as follows (see also Fig. 6):

Expansion (in.)	No. of Patients	Expansion (in.)	No. of Patients
0	8	1 1/4	9
1/4	8	1 1/2	22
1/2	23	1 3/4	10
3/4	24	2	21
1	25	Over 2	34

In 29 cases chest expansion was examined frequently over a period of 5 years or more; it improved, possibly as a result of therapy, in eighteen cases, was unchanged in five, and was worse in six.

Iritis and Iridocyclitis.—These ocular symptoms have long been recognized as more common in ankylosing spondylitis than in rheumatoid arthritis, but here again the length of the follow-up period will affect the results (Hart, 1951) and the incidence of 10 per cent. which is usually quoted, rises with an increase in the number of "observation years". Iritis may come on at any stage of the disease process and may also precede the spondylitis. In our series a history of iritis or iridocyclitis was forthcoming in 25 cases (13.5 per cent.). In one case iritis preceded spondylitis, in 24 cases it appeared at some time during the disease course, and in nine cases it occurred on more than one occasion. The incidence in our series will obviously rise with time.

Affection of the Sterno-Manubrial Joint.—This appears to be more common in ankylosing spondylitis than in rheumatoid arthritis. Savill (1951) found narrowing and eventual fusion of the joint in a high proportion of cases; 100 per cent. of spondylitics over the age of 35 showed abnormalities. Solovay and Gardner (1951) found fusion of the sterno-manubrial joint four times as common in ankylosing spondylitis (23 per cent. of cases) as in non-rheumatoid disease of the spine (5.5 per cent.). Françon and others (1953) state that though not uncommon, affection of this joint rarely gives

rise to symptoms; where it does so injections of local anaesthetic may help. In our experience mild symptoms arising from involvement of this joint are by no means uncommon, tenderness on pressure, swelling, and pain being not infrequent complaints; though rarely persisting for more than a few weeks such symptoms may cause considerable discomfort.

Bony Tenderness.—As already stated, this may be a major complaint in ankylosing spondylitis, the common sites being the ischial tuberosities, pelvic brim, greater trochanters, and occasionally over the sacrum and vertebral spinal processes. Tenderness over the sternum and ribs is not uncommon. Such tenderness may constitute a major complaint and warrant local deep x-ray therapy to the affected part; x-ray changes are characteristic (Figs 7 and 8a-d). Tenderness in the heel may also be a sign of



Fig. 7.—Pelvic osteopathic changes in ankylosing spondylitis. Note absence of change on inner surface of pelvis.

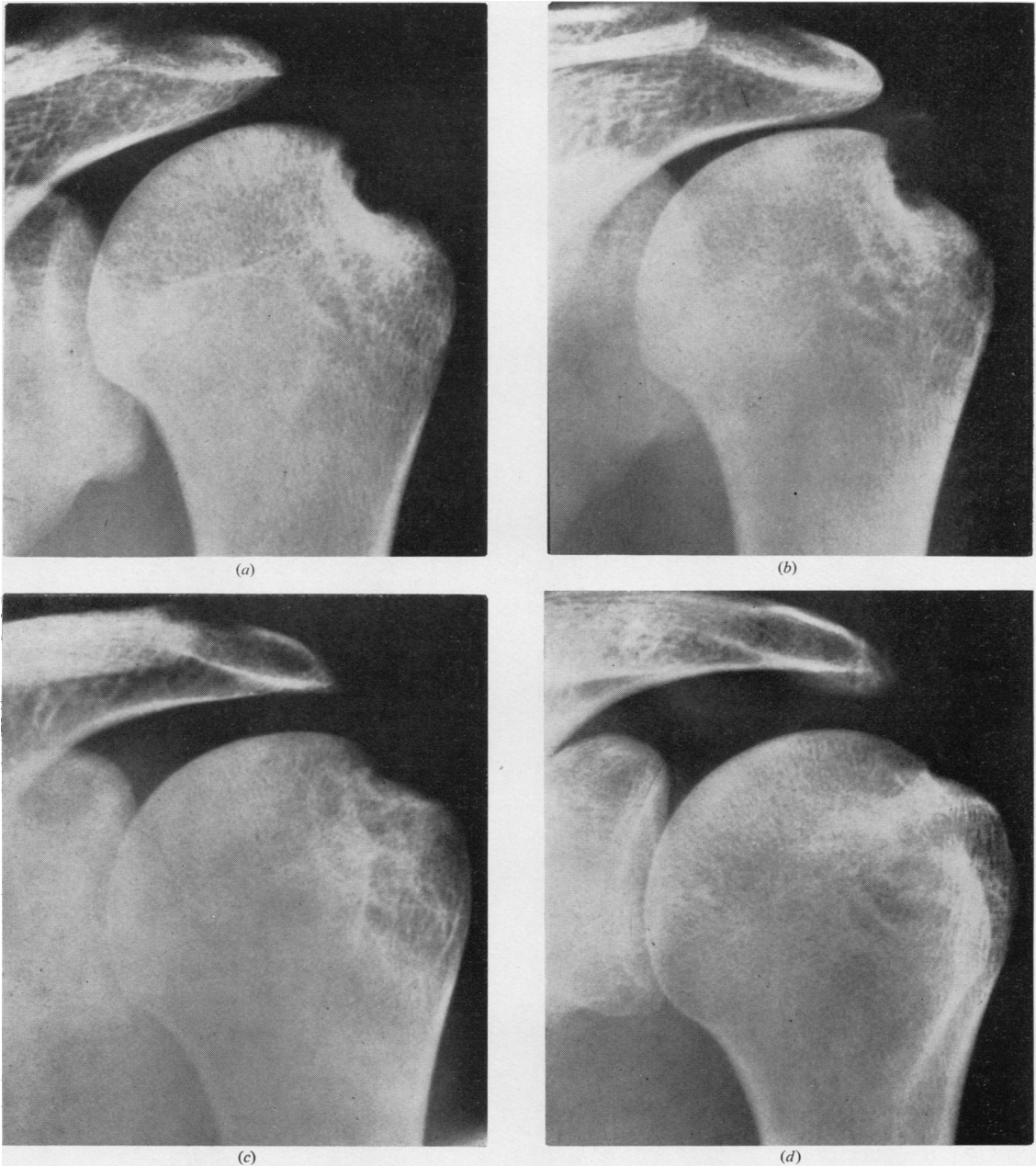


Fig. 8(a).—Bitten-out area in head of humerus in a classical spondylitic, who presented with acute pain in this area 24.9.49. All symptoms and signs disappeared in the subsequent five years, without local therapy.

Fig. 8(b). 18.4.50

Fig. 8(c). 5.6.51

Fig. 8(d). 3.5.54

ankylosing spondylitis; Davis and Blair (1950) state that males between the ages of 18 and 30 years with symptoms and signs of calcaneal periostitis or spurs should be suspected of having ankylosing spondylitis.

The diagnostic importance of these tender areas in bone is obvious. They are frequently misdiagnosed, commonly as tuberculous lesions, the resulting therapy—prolonged immobilization—producing unfortunate results. Ankylosing spondylitis

is a disease affecting bone as well as joint and ligament. It is an osteopathy as well as an arthritis; this is an important aspect of the disease which has often been overlooked.

Intercurrent Disease.—In these 184 patients, pulmonary tuberculosis with signs of activity was found in seven instances: in five the disease was parenchymal, whereas two presented with pleural effusion. Two further cases had quiescent disease and were merely kept under observation.

Dyspepsia of ulcer type was present without radiological abnormality in eight instances: six severe and two mild. Symptoms of a duodenal ulcer with a positive x-ray were found in seven instances: three had suffered from haematemesis or melaena, and one had perforated. A gastric ulcer was found on two occasions on barium meal examination, and also one case of carcinoma of the stomach. Melaena occurred once during phenylbutazone therapy, and twice during deep x-ray therapy, in both instances where a course of 2,000r was being administered to all ports. In all, therefore, 21 patients showed some evidence of peptic ulceration, three being associated with some form of therapy and 10 with radiological abnormality.

Summary

(1) In surveying the case histories of 184 patients with ankylosing spondylitis the onset of symptoms was found to have occurred before the age of 21 in 61 cases. Minor symptoms in peripheral joints may precede pain and stiffness in the back by months or years. In 24 cases (13 per cent.) pain and/or swelling of peripheral joints was the initial symptom. Four cases were misdiagnosed as rheumatic fever.

(2) Measurement of spinal movement is discussed. The "spondylometer" of Dr. W. F. Dunham has proved extremely useful.

(3) Of 184 patients, 88 had a chest expansion of 1 in. or less at nipple level at their initial attendance.

(4) Iridocyclitis was present in 25 instances (13.5 per cent.), active pulmonary tuberculous disease was noted in seven cases, and some evidence of peptic ulceration in 21 cases.

(5) The serum proteins and flocculation tests are recorded in 86 cases (see Appendix). The albumin was low, the globulin and fibrinogen were high, and the flocculation tests were positive in a proportion of cases. The changes were similar to those seen in rheumatoid arthritis but smaller in extent.

Our thanks are due to Dr. Peter Hansell of the Department of Medical Photography, Westminster Medical School.

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Spondylite ankylosante

RÉSUMÉ

(1) L'analyse des observations de 184 malades atteints de spondylite ankylosante montre que dans 61 cas les premiers symptômes ont débuté avant l'âge de 21 ans. Des mois et des années peuvent s'écouler entre le début des symptômes mineurs dans les articulations périphériques et l'apparition de la douleur et de la rigidité dorsale. Dans 24 cas (13%) la douleur et tuméfaction des articulations périphériques furent des symptômes initiaux. Chez quatre malades on fit le diagnostic erroné de rhumatisme articulaire aigu.

(2) On discute la mesure du mouvement vertébral. Le "spondylomètre" du Dr. W. F. Dunham s'est montré extrêmement utile.

(3) Au premier examen, l'ampliation thoracique au niveau du mamelon fut inférieure à 26 mm. chez 88 malades sur 184.

(4) L'iridocyclite fut présente dans 25 cas (13,5%), la tuberculose pulmonaire active dans 7 cas et dans 21 cas on observa des signes d'ulcération peptique.

(5) Les taux des protéines sériques et les résultats des réactions de flocculation furent notés dans 86 cas. Les taux d'albumine furent bas, ceux de la globuline et du fibrinogène élevés et les réactions de flocculation positives dans un certain nombre des cas. Les lésions furent similaires à celles observées dans l'arthrite rhumatismale mais de moindre envergure.

Espondilitis anquilosante

SUMARIO

(1) El análisis de 184 observaciones de enfermos con espondilitis anquilosante muestra que en 61 casos los primeros síntomas presentáronse antes de la edad de 21 años. Síntomas menores en articulaciones periféricas pueden preceder de meses y hasta de años el dolor y la rigidez dorsal. En 24 casos (13%) el dolor e hinchazón de las articulaciones periféricas fueron síntomas iniciales. En cuatro enfermos se hizo el diagnóstico erróneo de reumatismo poliarticular agudo.

(2) Se discute la medición del movimiento vertebral. El "espondilometro" del Dr. W. F. Dunham mostrose muy útil.

(3) Al examen inicial la ampliación torácica al nivel del pezón fué inferior a 26 mm. en 88 enfermos sobre los 184.

(4) La presencia de iridociclitis fué notada en 25 casos (13,5%), de tuberculosis pulmonar activa en 7 casos e indicios de ulceración péptica observáronse en 21 casos.

(5) Las cifras de las proteínas séricas y los resultados de las reacciones de flocculación fueron notados en 86 casos. Las cifras de albumina fueron bajas, las de globulina y de fibrinógeno altas y reacciones de flocculación positivas en un número determinado de los casos. Las alteraciones patológicas fueron similares a las observadas en la artritis reumatoide pero no tan extensas.

APPENDIX

LABORATORY INVESTIGATIONS

BY

N. F. MACLAGAN

Serum Proteins

These were estimated in some seventy patients with ankylosing spondylitis attending Westminster Hospital during the period 1950-52. 22 per cent. weight in volume sodium sulphate was used for precipitating the globulins and the standard micro-kjeldahl procedure was adopted. The results are shown in Table I; 21·2 per cent. of the cases had low albumin values while 33·2 per cent. had raised globulin values. The albumin/globulin ratio was below normal in 37·1 per cent. The plasma fibrinogen was also estimated in 38 patients with this condition, and 34·2 per cent. of these gave abnormally high results.

These figures differ fairly markedly from normal values, the deviation being in the same direction as that seen in rheumatoid arthritis. A parallel series of cases of rheumatoid arthritis investigated at the same time showed, however, a significantly greater deviation from the normal in the case of the albumin, globulin, and albumin/globulin ratio, but a rather smaller increase in the fibrinogen results (16 per cent. of cases abnormal).

The results as a whole indicate a considerable upset of protein metabolism in spondylitis of the

same type frequently associated with severe chronic infections.

Flocculation Tests

These were performed on a series of 86 cases with the results shown in Table II. All the flocculation tests tried showed a considerable percentage of positive results, the maximum occurring with the ammonium sulphate test (33 per cent.) and the minimum with the thymol turbidity and flocculation tests (11·6 and 12·8 per cent.). For technical reasons the serum colloidal gold test is to be preferred for demonstrating this type of change in rheumatic conditions; this gave 23·2 per cent. positive.

These figures, while showing a significant deviation from normal, are not so impressive as those obtained in rheumatoid arthritis, which in a parallel series of cases gave positive results ranging from 49·6 per cent. with the colloidal gold reaction to 26·7 per cent. with the thymol turbidity test.

As with the serum proteins, the changes are similar to those found in severe chronic infections and appear to be non-specific in character. It is possible that they depend partly on disturbance of liver function, but this is as yet unproved.

APPENDIX TABLE I
PLASMA PROTEINS IN SPONDYLITIS

Fraction	Normal Limits (g/100 ml.)	No. of Cases	Mean \pm SE (g./100 ml.)	Per cent. Abnormal
Albumin	4·0-5·5	66	4·321 \pm 0·074	21·2
Globulin	1·5-3·0	66	2·617 \pm 0·099	33·3
Fibrinogen	0·2-0·5	38	0·446 \pm 0·026	34·2
A/G Ratio	1·5-2·5	66	1·81 \pm 0·086	37·1

APPENDIX TABLE II
FLOCCULATION TESTS IN SPONDYLITIS

Test	Method	Normal Limits	No. of Cases	Mean \pm SE (units)	Per cent. Abnormal
Ammonium Sulphate ..	Huerga and others (1950)	0-2	18	2·25 \pm 0·038	33·3
Zinc Sulphate	Kunkel (1947)	0-4	65	3·190 \pm 0·29	16·9
Serum Colloidal Gold ..	Maclagan (1944a)	0	86	0·44 \pm 0·11	23·2
Thymol Turbidity	Maclagan (1944b)	0-2	86	1·48 \pm 0·13	11·6
Thymol Flocculation ..	Maclagan (1947)	0	86	0·15 \pm 0·45	12·8