JOINT DISEASE:
All the arthropathies

F. DUDLEY HART

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E. C. HUSKISSON
Senior Registrar
St. Bartholomew's Hospital, London

F. DUDLEY HART
Consultant Physician and Rheumatologist
Westminster Hospital, London

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INTRODUCTION

To make a diagnosis, the human brain functions as a computer, fitting the presence and absence of clinical features to the known characteristics of diseases. In order to make the correct diagnosis, it is therefore necessary to know as much about the rarest disease as the commonest, and this book aims to provide such information. It is not intended for reading, but for consultation when information is required. It is not a textbook, and having no pictures, makes no attempt to replace the experience of seeing patients which is essential to learning rheumatology.

THE INFORMATION IS ORGANIZED AS FOLLOWS:

Definition of disease X, including aetiology if known, and inheritance.

Incidence of arthritis in disease X; age and sex, type and stage of disease X in which arthritis is particularly seen; family history; persons specially predisposed.

Joints affected. How many? Which? How often? Symmetrical or asymmetrical? Migratory?

Symptoms, including speed of onset and precipitating factors.

Signs.

Course of the arthritis and of the underlying disease. If episodic, frequency, regularity, and duration of attacks. Effect of treatment.

Associations, *radiological findings*, and useful *laboratory tests.*

Treatments and *variants.*

Where possible, the exact frequency of any occurrence is given; this is based on a survey of the literature and modified by personal experience. Where it is not possible to give exact frequencies, the following approximations are used:

Usual	:	at least 90 per cent
Common	:	at least 70 per cent
Often	:	40–70 per cent
Occasional	:	10–40 per cent
Rare	:	less than 10 per cent

One or two recent references are given where possible, as a starting point for those seeking further information.

INCIDENCE OF ARTHROPATHIES

So that a correct diagnosis may be made, the frequency of different arthropathies must be known, as well as their clinical features and the clinical features of the case. For example, though rheumatoid arthritis is usually polyarticular, it is a much more likely cause of arthritis of a single joint than pigmented villonodular synovitis. The exact frequency of many diseases is not known, but in rheumatological practice in England one might expect to see the arthropathies in the following proportions:—

Several times each week:

Rheumatoid arthritis.
Osteo-arthrosis.
Normality with symptoms (psychogenic rheumatism or non-disease).
Frozen shoulder.
Traumatic and occupational arthropathies.

Several times each month:

Gout. Pyrophosphate arthropathy.
Psoriatic arthropathy.
Reiter's disease.
Ankylosing spondylitis.
Polymyalgia rheumatica.
Drug-induced arthropathies.

Several times each year:

Systemic lupus erythematosus.
Polyarteritis nodosa.
Polymyositis and dermatomyositis.
Septic arthritis.
Tuberculous arthritis.
Scleroderma.
Still's disease.
Palindromic rheumatism.
Pseudohypertrophic osteo-arthropathy and other arthropathies associated with malignant disease.
Neuropathic joints.
Shoulder-hand syndrome.
Ankylosing vertebral hyperostosis.
Arthritis associated with ulcerative colitis, rubella, sarcoidosis, and erythema nodosum.

The remainder are rare and should be diagnosed with caution. In other parts of the world, in general practice, and in other specialities such as paediatrics, the relative incidence of the arthropathies may be quite different.

CAUSES OF JOINT PAIN[1]

1. *Joint disease*: The arthropathies.
2. *Bone disease*: Fractures, primary or secondary tumours, osteochondritis, osteomyelitis, etc.
3. *Soft-tissue lesions*:
   Sprains and strains.
   Tenosynovitis.
   Overuse syndromes.
   'Soft-tissue rheumatism.'
   Direct trauma.
   Bursitis.
4. *Arthralgia*: Defined as joint pain in the absence of objective evidence of joint disease. This occurs in a variety of conditions, including:
   A. THE ARTHROPATHIES, either preceding the development of local signs or in some conditions in which there may be no local signs. Important examples are polymyalgia rheumatica and temporal arteritis, systemic lupus erythematosus and polyarteritis nodosa.
   B. INFECTIONS, particularly viral and rickettsial.
   Virus infections: influenza (25 per cent of cases), glandular fever, psittacosis, yellow fever, and sandfly fever.
   Rickettsial infections: all types of typhus.
   Bacterial infections: septicaemia, subacute bacterial endocarditis, typhoid and salmonella infections, and brucellosis.
   Spirochaetal infections: secondary syphilis, leptospirosis, and relapsing fever.
   Protozoal and metazoal infections: kala-azar and many other tropical, febrile conditions.
   C. DRUGS (*see* page 22), immunization, and allergies.
   D. PROTEIN ABNORMALITIES, e.g. mixed IgG IgM cryoglobulinaemia.[2]
5. *Referred pain* is particularly common in the shoulder joint when it may be due to:
   A. Abdominal conditions: liver abscess, subphrenic abscess, gall-bladder disease, peritonitis.
   B. Oesophageal conditions: hiatus hernia.
   C. Cardiac conditions: myocardial ischaemia.
   D. Neurological conditions: cervical cord or brachial plexus lesions.
   E. Pulmonary conditions: apical tumours, pleurisy, pneumothorax.
   Pain in the knee is often due to hip disease.
6. *Psychogenic*: Aches and pains are common in normal people, particularly women. Some have a tendency to joint pains from childhood which continue throughout life, often related to weather conditions (so-called 'rheumatism'). Joint pain may be a manifestation of psychological disturbance and rheumatism may become a source of complaint in anxious or neurotic patients.

REFERENCES

[1] HART, F. D. (1970), *Annals of Physical Medicine*, **10**, 257.
[2] MELTZER, M., and others (1966), *American Journal of Medicine*, **40**, 837.

HISTORY AND EXAMINATION

History

1. Age, sex, race, occupation.
2. Main complaint.
   a. When did it start?
   b. How did it start? (sudden, gradual, time of day).
   c. Precipitating factors.
   d. Which joints are affected?
   e. Morning stiffness.
   f. Pattern (continuous, episodic, migratory).
   g. If episodic, frequency, regularity, and duration of attacks.
   h. Associated symptoms.
3. Inquire particularly for skin disease, diarrhoea, venereal disease, drugs, and recent overseas visits.
4. Past medical history.
5. Family history. Gout. Diabetes.

Examination

1. *General appearance.*

   Pallor, pigmentation.
   Spot diagnoses: myxoedema, acromegaly.
   Posture.
   Gait.
2. *Examination of joints.*
   a. Overlying skin: colour, temperature, consistency (smooth, shiny, thick, thin, pitting), moistness.
   b. Swelling: articular or periarticular. Effusion (fluctuant), synovial thickening (soft non-fluctuant), or bony swelling (hard).
   c. Tenderness.
   d. Position (deformities): range of motion, crepitus.
   e. Periarticular soft tissues: Muscle wasting, tendon involvement, bursae, tender points.
3. *Complete physical examination essential.* Look particularly for tophi (ears or periarticular), nodules (ulnar surface of forearm), lymph-nodes, skin lesions, finger clubbing; record temperature. Routine examination of cardiovascular system, chest, abdomen, and central nervous system. Examine penis if suspicious of Reiter's disease.

JOINT PUNCTURE

INDICATIONS

1. Diagnostic, particularly suspicion of septic or infective arthritis and crystal deposition diseases (gout and pyrophosphate arthropathy).
2. For intra-articular injection of (*a*) steroids, particularly in chronic inflammatory arthritis for a single joint which is persistently painful despite rest and anti-inflammatory drugs or to aid mobilization of a joint, or (*b*) antibiotics, undesirable and unnecessary except for those antibiotics which are too toxic to give parenterally, e.g., polymyxin B.
3. To aspirate synovial fluid in septic arthritis; this is sometimes useful with very large effusions, haemarthroses, and pyrophosphate arthropathy.
4. For synovial biopsy, particularly for diagnosis of either rheumatoid arthritis or tuberculosis.

RISKS

1. Infection; about 1 in 10,000 if proper precautions are taken.
2. Destructive changes following repeated intra-articular steroid injection.
3. Inflammation due to injection of crystalline steroid preparations.

PRECAUTIONS

1. Strict asepsis.
2. Not more than three steroid injections in any joint.
3. Warn the patient (*a*) to report at once if the joint becomes more painful, and (*b*) to rest for at least 24 hours, and to avoid excessive weight-bearing for 3 weeks.

SITE: Descriptions such as 'above' and 'downwards' refer to the patient in position for injection and not to standard anatomical positions.

JOINT	PATIENT	NEEDLE ENTERS
Knee	Lying, knee slightly flexed	Medial side, below the patella, needle directed laterally and slightly downwards
Ankle	Lying	Just in front of, and medial to, medial malleolus, needle directed downwards and slightly laterally
Hip	Lying	2 cm. below the inguinal ligament just lateral to the femoral artery, needle directed downwards and slightly medially
Shoulder	Sitting; arm resting by side	Just below the coracoid process, needle directed posteriorly
Elbow	Sitting; elbow flexed to 90°	Behind and 1 cm. below the lateral epicondyle, needle directed medially, and slightly towards the hand
Wrist	Sitting; hand resting palm downwards	Just distal to ulnar styloid process, needle directed downwards
M.C.P. or *P.I.P.* joints	Sitting; hand resting palm downwards	Medial or lateral side of dorsal surface, needle directed downwards and laterally or medially

DOSE: Hydrocortisone or prednisolone, 25–50 mg. (1–2 ml.) for knee and other large joints, 12·5 mg. for small joints of hands and temporomandibular joint.

SYNOVIAL FLUID

After aspiration note volume, colour, clarity, turbidity, blood-staining, and viscosity. Divide as follows:

2 ml. in a tube containing EDTA for cell count and differential	At least 1 ml. in a sterile container for microscopy and culture. Ask for urgent Gram stain or special culture media if indicated	Remainder: examine a wet film for crystals under polarized light. Note presence or absence of clot formation after standing

RESULTS: The characteristics of synovial fluid depend upon the presence or absence of inflammation of synovium. *Non-inflammatory fluid* is clear, viscous, fails to clot on standing, and contains less than 1000 cells per c.mm., predominantly mononuclears. *Inflammatory fluid* is non-viscous, may clot, and contains an increased number of white blood-cells. These changes may be slight or gross, depending on the severity of the inflammatory process. Fluid with a high white-cell count is turbid and this does not necessarily mean that it is septic. The characteristics of inflammatory and non-inflammatory fluid are summarized in the following table. Synovial fluid characteristics in individual arthropathies are shown with other laboratory features of the conditions.

	NON-INFLAMMATORY	INFLAMMATORY		
	E.g., Osteo-arthrosis or Traumatic Arthritis	Rheumatoid Arthritis	Septic Arthritis	Gout or Pyrophosphate Arthropathy
Appearance	Clear	Often turbid	Turbid	Clear with flakes of fibrin
Colour	Yellow	Yellow/green	Brown/green	Yellow
Viscosity	High	Low	Low	Low
Clots?	No	Yes	Yes	Yes
Approx. W.B.C. (per mm.^{3})	1,000	30,000	100,000	10,000
Predominant cell	Mononuclears	Neutrophils	Neutrophils	Neutrophils
Crystals	No	No	No	Yes
Culture	Sterile	Sterile	Positive	Sterile

IDENTIFICATION OF CRYSTALS. Uric acid crystals are needle-shaped and strongly negatively birefringent (blue across the plane of the first-order red compensator); pyrophosphate crystals have square ends and are weakly positively birefringent (blue along the plane of the compensator).

SPECIAL TESTS

1. Complement levels are low in rheumatoid arthritis and high in Reiter's disease.
2. Latex tests parallel serum titres in patients with rheumatoid arthritis; there is a high incidence of false positives in other conditions and the test has little diagnostic value.

ABBREVIATIONS

ACTH:	Adrenocorticotrophin
ANF:	Antinuclear factor
ASO:	Anti-streptolysin O
cf.:	Compare
cm.:	Centimetres
c.mm.:	Cubic millimetre
C.N.S.:	Central nervous system
C.S.F.	Cerebrospinal fluid
CXR:	Chest X-ray
D.I.P.:	Distal inter-phalangeal
DNA:	Deoxyribonucleic acid
E.C.G.:	Electrocardiograph
E.M.G.:	Electromyograph
E.S.R.:	Erythrocyte sedimentation rate
F.:	Female
F.H.:	Family history
g.:	Grammes
I.M.:	Intramuscular
kg.:	Kilograms
LDH:	Lactic dehydrogenase
L.E.:	Lupus erythematosus
M.:	Male
M.C.P.:	Metacarpophalangeal
PBI:	Protein-bound iodine
P.I.P.:	Proximal interphalangeal
p.r.n.:	To be given when required
q.d.s.:	To be given four times daily
S.L.E.:	Systemic lupus erythematosus
t.d.s.:	To be given three times daily
T.P.I.:	*Treponema pallidum* immobilization test
W.B.C.:	White blood-count
W.R.:	Wassermann reaction
XR:	X-ray appearances
>:	Greater than
<:	Less than

ACROMEGALY

A pituitary adenoma causing chronic over-production of growth hormone which in adults results in gradual enlargement of the bones of the head, hands, and feet.

Incidence

F. 3 : 2.
Commonest onset age 20–40.
50 per cent have arthropathy.

Joints affected

Finger joints, spine, and knees often.
Hips, shoulders, ankles, wrists, elbows occasionally.
Polyarticular; symmetrical.

Symptoms

Pain in back and affected joints.
Intermittent swelling.
Stiffness unusual.

Signs

1. Enlargement of joints due to synovial thickening and bony outgrowth.
2. Crepitus.
3. Recurrent effusion common.
4. Increased mobility in early stages; later restriction of movement in severely affected peripheral joints, but not spine.

Course

Intermittent episodes of pain lasting weeks or months.
May progress slowly or rapidly to severe disabling arthropathy, resembling late stages of osteo-arthrosis.

Associated features

1. Bilateral carpal tunnel syndrome in 40 per cent.
   Improves after hypophysectomy.
2. Other features of acromegaly, typical facies, large hands, hypertension, etc.
   Look for visual field changes.

XR

1. 'Tufting' of distal phalanges.
2. Increased joint space.
3. Bony outgrowth around joints ('lipping') especially bases of distal phalanges ('hooks') and spine.
4. Small areas of calcification of joint capsule and cartilage.
5. Thickened widely spaced bony trabeculae.
6. Enlarged pituitary fossa.
   No erosions.

Laboratory

Confirm the diagnosis by insulin- and glucose-tolerance tests, growth hormone level.

Treatment

1. Analgesics.
2. Consider hypophysectomy but this has no effect on arthropathy.
3. Consider carpal tunnel decompression.
4. Surgery in advanced cases.

REFERENCES

BLUESTONE, R., and others (1971), *Annals of the Rheumatic Diseases*, **30**, 243.

KELLGREN, J. H., and others (1952), *Quarterly Journal of Medicine*, **21**, 405.

ACRO-OSTEOLYSIS

A group of conditions characterized by destruction and disappearance of bone. This may be primary or secondary to various joint diseases. Osteolysis of articulating surfaces occurs as a complication of rheumatoid arthritis, psoriatic arthropathy, avascular necrosis, and chronic infection. Osteolysis of distal phalanges of hands or feet occurs in scleroderma, Raynaud's disease, leprosy, and peripheral arterial obstruction. Primary osteolysis is of four types, all rare.

1. *Hereditary osteolysis*:[1] inherited as an autosomal dominant character.

Incidence
M. = F.
Onset about age 3.

Joints affected
Wrists and ankles.
Usually bilateral.

Symptoms
Joint pain and swelling.

Signs—course
In early stages, joints are warm, tender, and swollen with limitation of movement.
Later signs of inflammation disappear but stiffness and deformities appear over the years.
The condition stabilizes spontaneously in early adult life.

XR
Porosis of carpal and tarsal bones appears at about age 6, followed by localized destruction progressing to complete disappearance by age 30. There is concave deformity of the adjacent ends of long bones.

Laboratory
Normal E.S.R.
Latex test negative.

Treatment
Steroids give symptomatic relief but have no effect on the course of the disease. Splints may be useful to prevent deformity. Surgery to correct deformity when active stage is over.

2. *Osteolysis with nephropathy*:[2] a non-hereditary condition otherwise resembling hereditary osteolysis, beginning in childhood and usually affecting wrists and ankles, sometimes also hands, feet, and elbows. It is associated with progressive and ultimately fatal chronic glomerulonephritis.

3. *Gorham's disease* (Disappearing or Phantom Bone Disease):[3] a non-hereditary condition of both sexes, appearing at age 5–65 (peak 10–30). Any bone may be affected, usually at multiple sites, asymmetrical in distribution and often occurring after mild trauma. There is weakness and limitation of movement, but pain is mild or absent. The condition eventually stabilizes sometimes with a 'boneless' limb. Histology shows proliferation of thin-walled vessels.

4. *Distal osteolysis*:[4] inherited as an autosomal dominant character, appears between the ages of 8 and 32, and is characterized by progressive osteolysis of phalanges, metatarsals, or metacarpals with recurrent ulceration of the hands and feet, and sequestration of bone fragments. Fingers and toes may be lost, but the condition eventually heals.

REFERENCES

[1] SHURTLEFF, D. B. (1964), *Journal of the American Medical Association*, **188**, 363.
[2] TORG, J. S., and STEEL, H. S. (1968), *Journal of Bone and Joint Surgery*, **50A**, 1629.
[3] ABELL, J. M., and BADGLEY, C. E. (1961), *Journal of the American Medical Association*, **177**, 771.
[4] SCHINZ, H. R., and others (1951), *Roentgen—Diagnostics*, Vol. 1, p. 734. New York: Grune & Stratton.

AFRICAN HISTOPLASMOSIS

Infection with the fungus *Histoplasma duboisii*, found in West Africa. The source and mechanism of infection are unknown. Disease may be localized to the skin or disseminated, giving rise to granulomatous or suppurative lesions in bones, joints, liver, spleen, or lymph-nodes. In contrast to histoplasmosis (*see* p. 45) lung involvement is rare.

Incidence

M. 2 : 1.
Any age, peak 10–20.
About 20 per cent have arthritis.

Joints affected

Any joint or spine.
Usually monarticular.

Clinical features

Painful swollen joint.

Course

Chronic, may lead to joint destruction.

Associations

1. Skin lesions: painless papules which become nodules and may ulcerate.
2. Rarely hepatosplenomegaly, lymphadenopathy, or bone lesions.
3. Spinal involvement occasionally causes paraplegia.

XR

Well-defined osteolytic areas in bone adjacent to the joint.

Laboratory

Diagnosis is made by microscopical examination and culture of exudate from skin lesions or biopsy material from joint.

Treatment

Amphotericin B.

REFERENCE

COCKSHOTT, W. P., and LUCAS, A. O. (1964), *Quarterly Journal of Medicine*, 33, 223.

AGAMMAGLOBULINAEMIA

A rare disorder characterized by absence of gamma-globulin on electrophoresis and diminished ability to produce circulating antibodies in response to antigenic stimulation. There are two types: *congenital*, transmitted as a sex-linked, recessive character and occurring in male children, and *primary acquired*, non-hereditary and occurring in both sexes at any age.

Incidence

Age 2–10 (congenital type) or 10–50 (acquired type).
30 per cent have arthritis, less common with the acquired type.
Arthritis appears several years after onset of infections.

Joints affected

Polyarticular.
Commonly knees, P.I.P. and M.C.P. joints.
Often wrists, ankles, and elbows.
Occasionally shoulders and hips.
Often asymmetrical.

Symptoms

Pain, often mild, sometimes absent.
Swelling and stiffness.

Signs

Soft-tissue swelling, limitation of movement, tenderness, effusion, and synovial thickening.
Warmth or erythema rare.

Course

Mild; may be episodic or chronic with relapses and remissions.
Bacterial infections may be fatal.

Associations

1. Recurrent bacterial infections, e.g., meningitis, septicaemia, urinary infection.
2. Nodules, histologically resembling rheumatoid nodules found in 30 per cent of cases; occasionally bursitis or tenosynovitis.
3. Collagen disease, e.g., dermatomyositis or scleroderma.
4. Lymphoma.

XR

Usually normal.
Occasionally periarticular porosis.

Laboratory

Raised E.S.R.
Latex test negative.
Very low levels of immunoglobulins.
Synovial fluid: non-inflammatory.
Synovial biopsy: indistinguishable from rheumatoid arthritis.

Treatment

1. Anti-inflammatory drugs but not steroids.
2. Gamma-globulin.
3. Antibiotics for infections.

Variants

Septic arthritis.

REFERENCES

GOOD, R. A., and others (1957), *Journal of Laboratory and Clinical Medicine*, **49**, 343.

JANEWAY, C. A., and others (1956), *Transactions of the Association of American Physicians*, **69**, 93.

AMYLOIDOSIS

Amyloid infiltration of joint capsule, synovium, and tendons which gives rise to a polyarthritis resembling rheumatoid arthritis. This is usually associated with multiple myeloma but may also occur in primary amyloidosis with no apparent cause. Distinction is difficult since plasma cells are increased in the marrow in both conditions. There may be associated amyloid infiltration in other 'primary' sites.

Incidence

M. = F.
Age 30–70.
Rare complication of myeloma. May precede other manifestations by up to 3 years.

Joints affected

Polyarticular.
Commonly affects small joints of hands, wrists, and knees.
Often shoulders, elbows, ankles, hips, and feet.
Bilateral and symmetrical.

Symptoms

Pain, swelling, stiffness, and limitation of motion.
Morning stiffness.

Signs

Joints are swollen (effusion and synovial thickening) and tender but not red or warm.

Course

Arthritis may progress to flexion deformities resembling rheumatoid arthritis.
The condition is usually fatal within 5 years.

Associations

1. Bilateral carpal tunnel syndrome occurs in 50 per cent.
2. Nodules occur in 70 per cent, resembling those of rheumatoid arthritis but showing amyloid on biopsy.
3. Infiltration of amyloid elsewhere may cause heart failure or peripheral neuropathy.
4. Other features of myeloma, bone pain, anaemia, etc.

XR

Periarticular osteoporosis.
Look for lesions of myeloma particularly in skull and vertebrae.

Laboratory

Anaemia and raised E.S.R.
Latex test usually negative.
Proteins; paraprotein demonstrable with reduction in immunoglobulin levels.
Look for Bence-Jones proteinuria.
Synovial fluid: non-inflammatory.
Diagnosis confirmed by biopsy of synovium, carpal ligament, or nodules which show amyloid.

Treatment

Poor response to anti-inflammatory drugs.
Carpal tunnel decompression if necessary.
Otherwise symptomatic.

REFERENCE

GOLDBERG, A., and others (1964), *American Journal of Medicine*, 37, 653.

ANGIOKERATOMA CORPORIS DIFFUSUM (Fabry's Disease)

A rare hereditary disease, transmitted as a sex-linked, recessive character. Vacuolated cells containing glycolipid are seen in the walls of blood-vessels.

Incidence

Usually males; females rarely and mildly affected.
Onset in childhood.

Articular manifestations

1. Distal interphalangeal joints of the fingers are commonly affected by a degenerative arthropathy, bilateral and symmetrical. There is bony swelling and flexion deformity. The condition is mild.
2. Pain in the fingers or toes may be mistaken for arthritis, though it is not confined to the joints. It is often burning in character and exacerbated by heat or exertion. It may be episodic or constant with exacerbations associated with febrile episodes.
3. Avascular necrosis occurs rarely (*see* p. 11).

Course

Death is usual at age 30–50 from uraemia or heart failure.
Arthritis is a minor problem.

Associations

1. Skin lesions: clusters of dark-red lesions up to 4 mm. in diameter in a symmetrical distribution around the pelvis.
2. Premature cerebral vascular disease, hypertension, and myocardial infarction.
3. Proteinuria leading to uraemia.
4. Ankle oedema.
5. Corneal opacity.
6. Diarrhoea and bleeding piles.

XR

Degenerative changes in distal interphalangeal joints.

Laboratory

Raised E.S.R. in febrile episodes.
Skin biopsy confirms the diagnosis.
No biochemical abnormality.

Treatment

Symptomatic.

REFERENCE

WISE, D., and others (1962), *Quarterly Journal of Medicine*, **31**, 177.

ANKYLOSING SPONDYLITIS

A chronic condition of the spine and sacro-iliac joints in which early inflammatory changes are followed by progressive restriction of spinal movement associated with radiological calcification of spinal ligaments.

Incidence

Affects 0·4 per cent of males. M. 9 : 1.
Age at onset 15–30 but rarely earlier or later.
F.H. in 6 per cent.

Joints affected

Initially sacro-iliacs affected symmetrically and bilaterally with involvement of entire spine thereafter.
Shoulders and hips affected in 40 per cent; peripheral joints in 25 per cent, knees, commonest (15 per cent), also ankles (10 per cent), feet (5 per cent), wrists (5 per cent), and rarely fingers.
Ribs fuse on to vertebrae and transverse processes; sternomanubrial and sternoclavicular joints often affected.

Symptoms

Gradual onset of low backache and/or pain in both buttocks (pseudo-bilateral sciatica).
Morning stiffness.
15 per cent present with peripheral arthritis.

Signs

1. Restriction of all movements of spine.
2. Chest expansion reduced (< 5 cm.).
3. Bony points often tender (heels, sternum, ribs, pelvic brim, and ischial tuberosities).
4. Peripheral joints may be swollen and tender.

Course

Tendency for stiffness to increase in the first few years.
Subsequently chronic but tending to improve, though hip involvement may be disabling.

Associations

1. 1 per cent of cases develop aortic incompetence. 8 per cent have cardiac conduction defects.
2. Amyloidosis rare.
3. Iritis in 25 per cent.
4. Rarely atlanto-axial subluxation or fractures of rigid segments of the spine which may be fatal.
5. No nodules.

XR

Changes best seen in sacro-iliac joints and spine (D.10 to L.2).

1. *Pelvis.* Sacro-iliac joints usually abnormal; sclerosis; blurring of joint outline; later obliteration. Ischial tuberosities roughened and show 'whiskering' with periosteal elevations. Symphysis pubis may be blurred.
2. *Spine.* Syndesmophytes; longitudinal ligamentous calcification eventually producing 'bamboo' spine; lytic lesions appear particularly at the upper anterior corners of vertebral bodies; squaring of vertebrae.

Laboratory

E.S.R. raised in 80 per cent; often mild anaemia.
Latex test negative.
Synovial fluid: inflammatory; W.B.C. up to 20,000, mainly neutrophils.

Treatment

1. Exercise and exercises to maintain as full mobility as possible.
2. Phenylbutazone and indomethacin helpful in this programme. Night cover important. Indomethacin 100 mg. on retiring (by mouth or suppository). Not steroids.
3. Surgery only rarely needed, usually hip replacement. Spinal (wedge lumbar) osteotomy for marked kyphosis.
4. Radiotherapy in conservative dosage helpful but carries small risk of leukaemia.

Variants

1. A similar picture may occasionally be seen in Reiter's disease, psoriasis, ulcerative colitis, Crohn's disease, and rarely Whipple's disease.
2. Still's disease in boys may later lead to ankylosing spondylitis.
3. In women, the disease tends to be mild with limited spinal involvement. Pregnancy has no effect.

REFERENCES

CALABRO, J. J., and MALTZ, B. A. (1970), *New England Journal of Medicine*, **282**, 606.

HART, F. D. (1955), *Annals of the Rheumatic Diseases*, **14**, 77.

HART, F. D., and ROBINSON, K. (1959), *Annals of the Rheumatic Diseases*, **18**, 15.

WILKINSON, M., and BYWATERS, E. G. L. (1958), *Annals of the Rheumatic Diseases*, **17**, 209.

ANKYLOSING VERTEBRAL HYPEROSTOSIS (Baastrup Syndrome, Zuckergusswirbelsaüle)

A condition of the elderly in which there is exuberant bony outgrowth from the spine. The condition must be distinguished from ankylosing spondylitis (*see* p. 7) which occurs in young adults, and causes pain, restriction of spinal movement, often peripheral joint involvement, sacro-iliitis, and distinct X-ray changes. The X-ray appearance must be distinguished from paraspinal ossification in psoriatic arthopathy (*see* p. 85).

Incidence
Usually males.
Age 50+.

Joints affected
Spine (dorsal particularly).
No peripheral joint involvement.

Symptoms
Often none, sometimes stiffness.
No pain.

Signs
Normal or slight restriction of spinal movement.
No kyphosis.

Course
Benign: non-progressive.

Associations
Diabetes mellitus (often).

XR
Lateral views of spine show large bony outgrowths arising from the anterolateral aspect of the vertebral body.
They may extend upwards producing a 'candle-flame' appearance or downwards producing a 'dripping candle-wax' appearance.
They often join to form a continuous sheet of bone anterior to the vertebral bodies.
Sacro-iliac joints normal.

Laboratory
Normal E.S.R., calcium, phosphates, and alkaline phosphatase.
Test urine for sugar.

Treatment
Not required.
Maintain mobility.

REFERENCE

FORESTIER, J., and ROTES-QUEROL, J. (1950), *Annals of the Rheumatic Diseases*, **9**, 321.