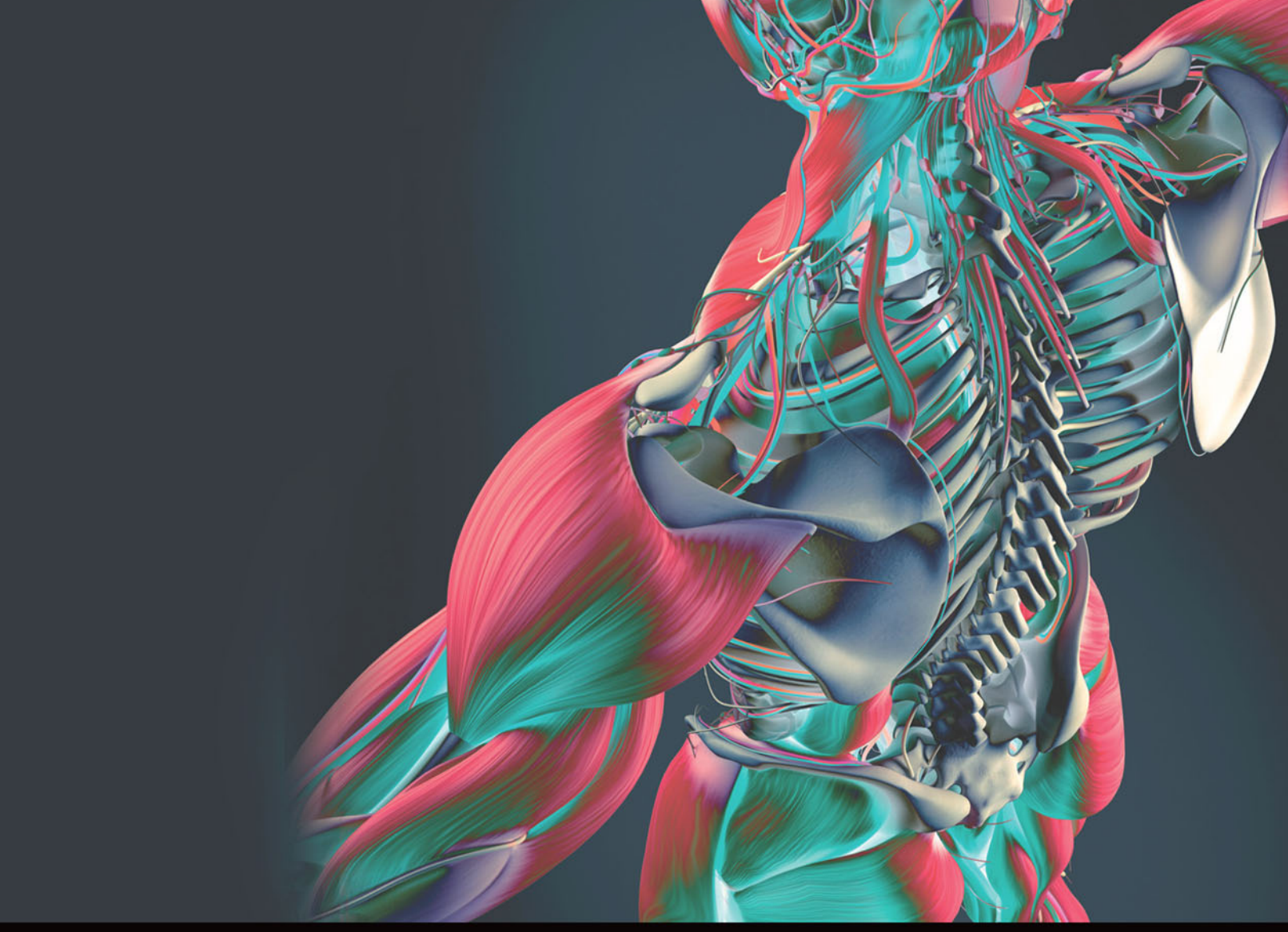


APLEY & SOLOMON'S

System of Orthopaedics and Trauma

Tenth Edition

EDITED BY

Ashley Blom, David Warwick, Michael R. Whitehouse

WITH VITALSOURCE® EBOOK

CRC Press
Taylor & Francis Group

Apley and Solomon's System of Orthopaedics and Trauma

Alan Graham Apley 1914–1996

Louis Solomon 1928–2014

Inspired teachers, wise mentors and joyful friends

Ashley W. Blom MBChB MD PhD FRCS FRCS (Tr&Orth)
Head of Translational Health Sciences
Bristol Medical School
University of Bristol
Bristol, UK

David Warwick MD BM FRCS FRCS(Orth) Eur Dip Hand Surg
Honorary Professor and Consultant Hand Surgeon
University of Southampton and University Hospital Southampton
Southampton, UK

Michael R. Whitehouse PhD MSc(Orth Eng) BSc(Hons)
PGCert(TLHE) FRCS(Tr&Orth) FHEA
Consultant Senior Lecturer in Trauma and Orthopaedics
University of Bristol
and
North Bristol NHS Trust
Musculoskeletal Research Unit
Southmead Hospital
Bristol, UK

Apley and Solomon's System of Orthopaedics and Trauma

Tenth Edition

CRC Press
Taylor & Francis Group
Boca Raton London New York

CRC Press is an imprint of the
Taylor & Francis Group, an **informa** business

CRC Press
Taylor & Francis Group
6000 Broken Sound Parkway NW, Suite 300
Boca Raton, FL 33487-2742

Printed on acid-free paper

International Standard Book Number-13: 978-1-4987-5167-4 (Pack – Book and eBook)
International Standard Book Number-13: 978-1-4987-5177-3 (Paperback; restricted territorial availability)

Library of Congress Cataloging-in-Publication Data

Names: Blom, Ashley, editor. | Warwick, David, 1962- editor. | Whitehouse, Michael (Michael R.), editor. | Preceded by (work): Solomon, Louis. Apley's system of orthopaedics and fractures.
Title: Apley & Solomon's system of orthopaedics and trauma / [edited by] Ashley Blom, David Warwick, Michael Whitehouse.
Other titles: Apley and Solomon's system of orthopaedics and trauma | System of orthopaedics and trauma.
Description: Tenth edition. | Boca Raton : CRC Press, [2017] | Preceded by Apley's system of orthopaedics and fractures / Louis Solomon, David Warwick, Selvadurai Nayagam. 9th ed. 2010.
Identifiers: LCCN 2016059350 (print) | LCCN 2016059955 (ebook) | ISBN 9781498751674 (hardback bundle : alk. paper) | ISBN 9781498751773 (pbk. : alk. paper) | ISBN 9781498751711 (eBook VitalSource) | ISBN 9781498751704 (eBook PDF).
Subjects: | MESH: Orthopedic Procedures | Musculoskeletal System--injuries | Fracture Fixation—methods.
Classification: LCC RD731 (print) | LCC RD731 (ebook) | NLM WE 168 | DDC 616.7--dc23
LC record available at https://lccn.loc.gov/2016059350

Visit the Taylor & Francis Web site at
http://www.taylorandfrancis.com

and the CRC Press Web site at
http://www.crcpress.com

DEDICATION

To Louis
from your friends and colleagues on behalf of the thousands of patients
who have benefitted from your lifetime's work

Taylor & Francis
Taylor & Francis Group
http://taylorandfrancis.com

CONTENTS

CONTRIBUTORS

Yves Acklin MD DMedSc EBSQ Trauma
Consultant Trauma and Orthopaedic Surgeon and Medical Fellow
Kantonsspital Graubünden
Chur, Switzerland
and
AO Research Institute
Davos, Switzerland

Richard P. Baker MD MSc FRCS(Tr&Orth)
Consultant Trauma and Orthopaedic Surgeon
North Bristol NHS Trust
Department of Trauma and Orthopaedics
Avon Orthopaedic Centre, Southmead Hospital
Bristol, UK

Ashley W. Blom MBChB MD PhD FRCS FRCS (Tr&Orth)
Head of Translational Health Sciences
Bristol Medical School
University of Bristol
Bristol, UK

Nick Bottomley DPhil FRCS(Orth)
Consultant Knee Surgeon
Nuffield Orthopaedic Centre
Oxford, UK

Gavin Bowyer MA FRCS
Consultant Orthopaedic Surgeon
Spire Hospital
Southampton, UK

Emma M. Clark MB BS MSc PhD FRCP
Musculoskeletal Research Unit
University of Bristol
Bristol, UK

Andrew Cole BSc(Hons) MBBS FRCS (Tr&Orth)
University Hospital Southampton NHS Foundation Trust
Southampton, UK

Paul Creamer MD FRCP
Consultant Rheumatologist
North Bristol NHS Trust
Bristol, UK

Gorav Datta MD FRCS(Tr&Orth)
Consultant Orthopaedic Surgeon
Honorary Senior Clinical Lecturer
University Hospital Southampton NHS Foundation Trust
Southampton, UK

Paul Dieppe BSc MD FRCP FFPH
Emeritus Professor of Health and Wellbeing
University of Exeter Medical School
St Luke's Campus
Exeter, UK

Robert Dunn MBChB(UCT) MMed(Orth) FCS(SA)Orth
Consultant Spine and Orthopaedic Surgeon
Pieter Moll and Nuffield Chair of Orthopaedic Surgery, University of Cape Town
Head, Division of Orthopaedic Surgery
Head, Orthopaedic Spinal Services, Groote Schuur Hospital
Spine Deformity Service Red Cross Children's Hospital
Cape Town, South Africa

Roderick Dunn MB BS DMCC FRCS(Plast)
Consultant Plastic Reconstructive and Hand Surgeon
Odstock Centre for Plastic Surgery and Burns, Salisbury Hospital
Salisbury, UK

Deborah Eastwood MB FRCS
Consultant Paediatric Orthopaedic Surgeon
Great Ormond St Hospital for Children and the Royal National Orthopaedic Hospital
London, UK

Christopher Edwards BSc MBBS MD FRCP
Professor and Consultant Rheumatologist
NIHR Wellcome Trust Clinical Research Facility
University Hospital Southampton NHS Foundation Trust
Southampton, UK

Michael Fox FRCS(Tr&Orth)
Consultant Surgeon in Peripheral Nerve Injury
The Royal National Orthopaedic Hospital
Stanmore, UK

Martin Gargan MA(Oxon) FRCS FRCS(Tr&Orth)
Head, Division of Paediatric Orthopaedics
Harold and Bernice Groves Chair in Orthopaedics
Hospital for Sick Children
Toronto, Canada

Enrique Gómez-Barrena MD, PhD
University Chair of Orthopaedic Surgery and Traumatology
Facultad de Medicina
Universidad Autónoma de Madrid
Madrid, Spain
and
Orthopaedic Surgeon
Head of the Lower Limb Reconstructive Surgery Unit at Hospital de Traumatología
La Paz University Hospital
Madrid, Spain

Boyko Gueorguiev PhD
Program Leader Biomedical Development
AO Research Institute
Davos, Switzerland

Nick Howells MSc MD FRCS (T&O)
Consultant Trauma and Orthopaedic Surgeon
Avon Orthopaedic Centre
Bristol, UK

William Jackson FRCS(Orth)
Consultant Knee Surgeon
Nuffield Orthopaedic Centre
Oxford, UK

Max Jonas MBBS FRCA FFICM
Consultant and Senior Lecturer
University Hospital Southampton and University of Southampton
Southampton, UK

Stephen A. Jones BSc(Hons) MBBCh MRCS(Eng) MSc(Orth Eng) FRCS(Orth)
Senior Lecturer in Orthopaedics
Cardiff University
and
Consultant Orthopaedic Surgeon
Cardiff & Vale University Health Board
Cardiff, UK

Dimitrios Kassimos MD MSc
Consultant Rheumatologist
General Military Hospital of Athens
Athens, Greece

Ryan Katchky BEng MD, FRCSC
Clinical Fellow
Hospital for Sick Children
Toronto, Canada

Nicholas Kruger BSc MBChB FRCS(Ed) FCSOrth(SA)
Consultant Orthopaedic and Spinal Surgeon
Head of Acute Spinal Cord Injury Unit
University of Cape Town Student Orthopaedic Training Coordinator
Groote Schuur Hospital
Cape Town, South Africa

Nuno Lança MD FEBOT
Orthopaedic Surgeon
Clinical Assistant of Hospital de Santa Maria Orthopaedic Department, Spinal Unit
Clinical Assistant of Hospital CUF Descobertas, Orthopaedic Department, Spinal Unit
Lisbon, Portugal

Jason Peter Mansell BSc(Hons) PhD
Senior Lecturer in Bone Biology
University of the West of England
Bristol, UK

Jorge Mineiro MD PhD FRCSEd
Orthopaedic Surgeon
Professor of Orthopaedics and Traumatology
Clinical Director of Hospital CUF Descobertas
Head of Hospital CUF Descobertas Orthopaedic Department
Head of Hospital CUF Descobertas Spinal Unit
Lisbon, Portugal

Fergal Monsell MSc PhD
Consultant Paediatric Orthopaedic Surgeon
Bristol Children's Hospital
Bristol, UK

Fintan T. Moriarty PhD
Senior Scientist Musculoskeletal Infection
AO Research Institute
Davos, Switzerland

Michael Parry BSc(Hons) MBChB PGCME MD FRCS(Tr&Orth)
Consultant Orthopaedic Surgeon
Department of Orthopaedic Oncology
Royal Orthopaedic Hospital
Birmingham, UK

Andrew Price DPhil FRCS(Orth)
Professor of Orthopaedic Surgery
Nuffield Department of Orthopaedics, Rheumatology and Musculoskeletal Science
Nuffield Orthopaedic Centre
Oxford, UK

R. Geoff Richards PhD
Director
AO Research Institute
Davos, Switzerland

H. Srinivasan MB BS FRCS FRCS Ed DSc (Hon)
Formerly Senior Orthopaedic Surgeon
Central Leprosy Teaching & Research Institute
Chengalpattu (Tamil Nadu), India;
Director Central JALMA Institute for Leprosy (ICMR)
Agra (UP), India
and
Editor, Indian Journal of Leprosy

Jonathan Daniel Stevenson MBChB BMedSci FRCS(Tr&Orth)
Consultant Orthopaedic Surgeon
Department of Orthopaedic Oncology
Royal Orthopaedic Hospital
Birmingham, UK

Martin Stoddart MPhil PhD
Principal Scientist
AO Research Institute
Davos, Switzerland
and
Albert-Ludwigs University
Freiburg, Germany

David Sutton BM DA FRCA
Consultant Anaesthetist
University Hospital Southampton NHS Foundation Trust
Southampton, UK

Joanna Thomas MBBS MSc FRCS(Tr&Orth)
Consultant Orthopaedic Surgeon
University Hospital Southampton NHS Foundation Trust
Southampton, UK

Jonathan H. Tobias MA MD PhD FRCP
Musculoskeletal Research Unit
University of Bristol
Bristol, UK

James Turner FRCS (Tr&Orth)
Consultant Orthopaedic Surgeon
CURE Ethiopia Children's Hospital
Addis Ababa, Ethiopia

Michael G. Uglow MBBS FRCS(Eng) FRCS(Glas) FRCS(Tr&Orth)
Consultant Orthopaedic Surgeon
University of Southampton and University Hospital Southampton NHS Foundation Trust
Southampton, UK

Charles J Wakeley BSc MBBS FRCS FRCSed FRCR
Consultant Radiologist, Department of Radiology
University Hospitals Bristol NHS Foundation Trust
Bristol, UK

David Warwick MD BM FRCS FRCS(Orth) Eur Dip Hand Surg
Honorary Professor and Consultant Hand Surgeon
University of Southampton and University Hospital Southampton
Southampton, UK

Adam C. Watts MBBS BSc FRCS(Tr&Orth)
Consultant Upper Limb Surgeon
Wrightington Hospital
Visiting Professor, University of Manchester
Manchester, UK

Michael Richard Whitehouse PhD MSc(Orth Eng) BSc(Hons) PGCert(TLHE) FRCS(Tr&Orth) FHEA
Consultant Senior Lecturer in Trauma and Orthopaedics
University of Bristol
and
North Bristol NHS Trust
Musculoskeletal Research Unit
Southmead Hospital
Bristol, UK

Taylor & Francis
Taylor & Francis Group
http://taylorandfrancis.com

PREFACE

Orthopaedics in a changing world

Since Alan Apley published the first edition of this book the world has changed considerably and so has the practice of orthopaedic surgery. In 1959, hip replacement was rare and had high failure rates, knee replacement and arthroscopy did not exist and fractures were primarily treated in traction.

The last edition of this book commented on the projected impact of the HIV/AIDS epidemic. The epidemic has largely been brought under control, with effective treatment resulting in normal life expectancy for sufferers. However, in untreated individuals, the incidence of secondary infection such as tuberculosis is high and the prognosis is still dire. It is interesting and encouraging to note that both the National Joint Registry for England and Wales and the Malawian Joint Registry have shown that hip replacement is an effective treatment for patients who have multimorbidity which includes AIDS with no increased risk of early postoperative mortality compared with patients who do not have AIDS.

Over the lifetime of this book many treatments have been invented, extensively used, found to be ineffective or suboptimal and subsequently have declined dramatically in popularity. Examples of this include arthroscopic debridement for knee osteoarthritis, metal-on-metal hip replacement and excision arthroplasty of the distal ulna. It is important that we continue to challenge the efficacy of existing and novel treatments. In a world of increasing global need orthopaedics has to be proven to be efficacious and cost-effective.

Since 1959, the world's population has more than doubled to over 7 billion people and has aged considerably. Life expectancy at birth is now 80 years in Europe and 74 years in Asia. There are still marked disparities – for instance Japan has a life expectancy at birth of 83 years compared to 57 years in South Africa – but these differences are narrowing. It is projected that by 2050 4% of the world's population (but 16% of Japan's population) will be over 80 years of age. Between 2010 and 2050 the proportion of the population aged over 65 years will double in most countries, and it is predicted to increase from 5% to 11% in South Africa, 5% to 13% in India and 17% to 36% in Spain.

Orthopaedics remains as relevant a speciality as ever, treating a large burden of the world's morbidity. However, the nature of care has changed, with a much lower burden of chronic musculoskeletal infections today and a steeply rising incidence of joint replacement for primarily degenerative conditions. The World Health Organization estimates that 10% of men and 18% of women aged over 60 years have symptomatic osteoarthritis. Total knee and total hip replacement are now the second and third commonest elective operative procedures performed in developed countries. For example, in England and Wales, which have a combined population of approximately 55 million people, over 170 000 hip and knee replacements are performed annually. The provision of arthroplasty varies greatly, with 226 knee replacements per 100 000 population performed annually in the United States of America compared to only 3 per 100 000 population in neighbouring Mexico. Increasingly the outcomes of common procedures, such as arthroplasty and fractured neck of femur fixation, are being monitored by national registries in a wide range of countries and healthcare settings. It is heartening that even low-income countries such as Malawi have established implant registries which are providing clinically important data. As the prevalence of infectious diseases declines in low-income countries and people live longer, more health resources will be spent on treating long-term conditions of the elderly such as osteoarthritis.

Accidents and emergencies still represent a major healthcare burden. Over 1.25 million people die worldwide annually as a result of road traffic accidents. The majority of these occur in Asia. Millions more are seriously injured. Injuries from road traffic accidents are the third largest cause of morbidity among adult males. Orthopaedic care remains of paramount importance for effectively and quickly returning patients as closely as possible to their pre-injury state and thereby allowing them to participate fully in society.

The provision of health care and resources varies considerably between countries: Greece has 6.3 doctors per 1000 population, South Africa has 0.8 and India only 0.7. While the number of doctors practising in some countries has remained relatively static, in Australia and the United Kingdom there has been an increase of over 50% in the number of registered doctors in the past decade. Part of this is due to migration of doctors, which may exacerbate shortages in low-income countries. More than 40 000 foreign-trained doctors, including an author of this preface, work in the United Kingdom, nearly half of whom come from India and Pakistan. In Israel, New Zealand, Norway and Ireland over a third of practising doctors are foreign-trained. Movement of doctors between countries promotes the spread of ideas and innovation and improves training. However, there is a natural gravitation of expertise towards countries that offer higher remuneration and better working conditions at the expense of low- and middle-income countries. The United States of America spends $8713 per capita on health care, while China spend $649 and India $215.

With rapidly increasing per capita GDP in countries such as China and India, the demographics of health care will change markedly over the next decade. The relative need to treat infection and injury will hopefully decline, but this will inevitably be coupled with an increase in treatments for longer-term musculoskeletal conditions.

Ashley W. Blom
David Warwick
Michael R. Whitehouse
Bristol and Southampton, 2017

Data are publically available from the OECD at:
http://www.oecd-ilibrary.org/social-issues-migration-health/health-at-a-glance_19991312#

PREFACE TO THE NINTH EDITION

When Alan Apley produced the first edition of his System of Orthopaedics and Fractures 50 years ago he saw it as an aid to accompany the courses that he conducted for aspiring surgeons who were preparing for the FRCS exams. With characteristic humour, he called the book 'a prophylactic against writer's cramp'. Pictures were unnecessary: if you had any sense (and were quick enough to get on the heavily oversubscribed Apley Course), you would be treated to an unforgettable display of clinical signs by one of the most gifted of teachers.

You also learnt how to elicit those signs by using a methodical clinical approach – the Apley System. The Fellowship exam was heavily weighted towards clinical skills. Miss an important sign or stumble over how to examine a knee or a finger and you could fail outright. What Apley taught you was how to order the steps in physical examination in a way that could be applied to every part of the musculoskeletal system. 'Look, Feel, Move' was the mantra. He liked to say that he had a preference for four-letter words. And always in that order! Deviate from the System by grasping a patient's leg before you look at it minutely, or by testing the movements in a joint before you feel its contours and establish the exact site of tenderness and you risked becoming an unwilling participant in a theatrical comedy.

Much has changed since then. With each new edition the System has been expanded to accommodate new tests and physical manoeuvres developed in the tide of super-specialization. Laboratory investigations have become more important and imaging techniques have advanced out of all recognition. Clinical classifications have sprung up and attempts are now made to find a numerical slot for every imaginable fracture. No medical textbook is complete without its 'basic science' component, and advances are so rapid that changes become necessary within the period of writing a single edition. The present volume is no exception: new bits were still being added right up to the time of proofreading.

For all that, we have retained the familiar structure of the Apley System. As in earlier editions, the book is divided into three sections: General Orthopaedics, covering the main types of musculoskeletal disorder; Regional Orthopaedics, where we engage with these disorders in specific parts of the body; and thirdly Fractures and Joint Injuries. In a major departure from previous editions, we have enlisted the help of colleagues who have particular experience of conditions with which we as principal authors are less familiar. Their contributions are gratefully acknowledged. Even here, though, we have sought their permission to 'edit' their material into the Apley mould so that the book still has the sound and 'feel' of a single authorial voice.

For the second edition of the book, in 1963, Apley added a new chapter: 'The Management of Major Accidents'. Typically frank, he described the current arrangements for dealing with serious accidents as 'woefully inadequate' and offered suggestions based on the government's Interim Report on Accident Services in Great Britain and Ireland (1961). There has been a vast improvement since then and the number of road accident deaths today is half of what it was in the 1960s (Department of Transport statistics). So important is this subject that the relevant section has now been rewritten by two highly experienced Emergency and Intensive Care Physicians and is by far the longest chapter in the present edition.

Elsewhere the text has been brought completely up to date and new pictures have been added. In most cases the illustrations appear as composites – a series of images that tell a story rather than a single 'typical' picture at one moment in the development of some disorder. At the beginning of each Regional chapter, in a run of pictures we show the method of examining that region: where to stand, how to confront the patient and where to place our hands. For the experienced reader this may seem like old hat; but then we have designed this book for orthopaedic surgeons of all ages and all levels of experience. We all have something to learn from each other.

As before, operations are described only in outline, emphasizing the principles that govern the choice of treatment, the indications for surgery, the design of the operation, its known complications and the likely outcome. Technical procedures are learnt in

simulation courses and, ultimately, in the operating theatre. Written instructions can only ever be a guide. Drawings are usually too idealized and 'in theatre' photographs are usually intelligible only to someone who has already performed that operation. Textbooks that grapple with these impediments tend to run to several volumes.

The emphasis throughout is on clinical orthopaedics. We acknowledge the value of a more academic approach that starts with embryology, anatomy, biomechanics, molecular biology, physiology and pathology before introducing any patient to the reader. Instead we have chosen to present these 'basic' subjects in small portions where they are relevant to the clinical disorder under discussion: bone growth and metabolism in the chapter on metabolic bone disease, genetics in the chapter on osteodystrophies, and so forth.

In the preface to the last edition we admitted our doubts about the value of exhaustive lists of references at the end of each chapter. We are even more divided about this now, what with the plethora of 'search engines' that have come to dominate the internet. We can merely bow our heads and say we still have those doubts and have given references only where it seems appropriate to acknowledge where an old idea started or where something new is being said that might at first sight be questioned.

More than ever we are aware that there is a dwindling number of orthopaedic surgeons who grew up in the Apley era, even fewer who experienced his thrilling teaching displays, and fewer still who worked with him. Wherever they are, we trust that they will recognize the Apley flavour in this new edition. Our chief concern, however, is for the new readers who – we hope – will glean something that helps them become the next generation of teachers and mentors.

LS
SN
DJW

ACKNOWLEDGEMENTS

This textbook is an iterative process and for this current edition new authors have been asked to revise and refresh the existing text. The editors and new authors thoroughly acknowledge the contribution of those who have gone before them, much of whose work remains in this updated text.

Chapter 2, *Infection*, contains some material from 'Infection' by Louis Solomon, H. Srinivasan, Surendar Tuli & Shunmugam Govender. The material has been revised and updated by the current author.

Chapter 4, *Crystal deposition disorders*, contains some material from 'Crystal deposition disorders' by Louis Solomon. The material has been revised and updated by the current authors.

Chapter 5, *Osteoarthritis*, contains some material from 'Osteoarthritis and related disorders' by Louis Solomon. The material has been revised and updated by the current authors.

Chapter 6, *Osteonecrosis and osteochondritis*, contains some material from 'Osteonecrosis and osteochondritis' by Louis Solomon. The material has been revised and updated by the current authors.

Chapter 7, *Metabolic and endocrine bone disorders*, contains some material from 'Metabolic and endocrine bone disorders' by Louis Solomon. The material has been revised and updated by the current authors.

Chapter 8, *Genetic disorders, skeletal dysplasias and malformations*, contains some material from 'Genetic disorders, skeletal dysplasias and malformations' by Louis Solomon & Deborah Eastwood. The material has been revised and updated by the current authors.

Chapter 9, *Tumours*, contains some material from 'Tumours' by Will Aston, Timothy Briggs & Louis Solomon. The material has been revised and updated by the current authors.

Chapter 10, *Neuromuscular disorders*, contains some material from 'Neuromuscular disorders' by Deborah Eastwood, Thomas Staunton & Louis Solomon. The material has been revised and updated by the current author.

Chapter 11, *Peripheral nerve disorders*, contains some material from 'Peripheral nerve injuries' by David Warwick, H. Srinivasan & Louis Solomon. The material has been revised and updated by the new contributor Michael Fox.

Chapter 12, *Principles of orthopaedic operations*, contains some material from 'Principles of orthopaedic operations' by Selvadurai Nyagam & David Warwick. The material has been revised and updated by the current authors.

Chapter 13, *The shoulder and pectoral girdle*, contains some material from 'The shoulder and pectoral girdle' by Andrew Cole & Paul Pavlou. The material has been revised and updated by Andrew Cole.

Chapter 14, *The elbow*, contains some material from 'The elbow and forearm' by David Warwick. The material has been revised and updated by the new contributor Adam Watts.

Chapter 16, *The hand*, contains some material from 'The hand' by David Warwick & Roderick Dunn. The material has been revised and updated by the same authors.

Chapter 17, *The neck*, contains some material from 'The neck' by Stephen Eisenstein & Louis Solomon. The material has been revised and updated by the current authors.

Chapter 18, *The back*, contains some material from 'The back' by Stephen Eisenstein, Surendar Tuli & Shunmugam Govender. The material has been revised and updated by the current authors.

Chapter 19, *The hip*, contains some material from 'The hip' by Louis Solomon, Reinhold Ganz, Michael Leunig, Fergal Monsell & Ian Learmonth. The material has been revised and updated by the current authors.

Chapter 20, *The knee*, contains some material from 'The knee' by Louis Solomon & Theo Karachalios. The material has been revised and updated by the current authors.

Chapter 23, *Principles of fractures*, contains some material from 'Principles of fractures' by Selvadurai Nayagam. The material has been revised and updated by the current authors.

Chapter 24, *Injuries of the shoulder and upper arm*, contains some material from 'Injuries of the shoulder, upper arm & elbow' by Andrew Cole, Paul Pavlou & David Warwick. The material has been revised and updated by Andrew Cole.

Chapter 25, *Injuries of the elbow and forearm*, contains some material from 'Injuries of the shoulder, upper arm & elbow' by Andrew Cole, Paul Pavlou & David Warwick, and some material from 'Injuries of the forearm and wrist' by David Warwick. The material has been revised and updated by the new contributors Adam Watts, Mike Uglow and Joanna Thomas.

Chapter 26 The wrist Contains some material from 'Injuries of the Forearm and Wrist' by David Warwick with updates from the new contributors Adam Watts, Mike Uglow and Joanna Thomas.

Chapter 28, *Injuries of the spine*, contains some material from 'Injuries of the spine' by Stephen Eisenstein & Wagih El Masry. The material has been revised and updated by the current authors.

Chapter 29, *Injuries of the pelvis*, contains some material from 'Injuries of the pelvis' by Louis Solomon. The material has been revised and updated by the current author.

Chapter 30, *Injuries of the hip and femur*, contains some material from 'Injuries of the hip and femur' by Selvadurai Nayagam. The material has been revised and updated by the current authors.

Chapter 31, *Injuries of the knee and leg*, contains some material from 'Injuries of the knee and leg' by Selvadurai Nayagam. The material has been revised and updated by the current author.

LIST OF ABBREVIATIONS USED

AAS	atlantoaxial subluxation
ABC	aneurysmal bone cyst
ABPI	ankle brachial pressure index
ACA	angulation correction axis
ACDF	anterior cervical discectomy and fusion
ACE	angiotensin-converting enzyme
ACEI	angiotensin-converting enzyme inhibitor
ACL	anterior cruciate ligament
ACLR	anterior cruciate ligament reconstruction
ACPA	anti-citrullinated peptide antibodies
ACTH	adrenocorticotropic hormone
ADH	antidiuretic hormone
ADHD	attention deficit hyperactivity disorder
ADHR	autosomal dominant hypophosphataemic rickets
ADI	atlantodental interval
ADL	activity of daily living
AFO	ankle–foot orthosis
AFP	alpha-fetoprotein
AIDP	acute inflammatory demyelinating polyneuropathy
AIDS	acquired immune deficiency syndrome
AJCC	American Joint Committee on Cancer
AL	anterolateral
ALI	acute lung injury
ALIF	anterior lumbar interbody fusion
ALP	alkaline phosphatase
ALS	amyotrophic lateral sclerosis
AM	anteromedial
AMC	arthrogryposis multiplex congenita
ANA	antinuclear antibodies
anti-CCP	anti-cyclic citrullinated peptide antibodies
AO/ASIF	Arbeitsgemeinschaft für Osteosynthesefragen/Association for the Study of Internal Fixation
AP	anteroposterior
APACHE	Acute Physiology and Chronic Health Evaluation (model)
APC	antigen-presenting cell *and* anteroposterior compression (injuries)
ARCO	Association Research Circulation Osseous
ARDS	acute respiratory distress syndrome
ARHR	autosomal recessive hypophosphatemic rickets
ARM	awareness, recognition, management
ARMD	adverse reaction to metal debris
AS	ankylosing spondylitis
ASCT	autologous stem-cell transplantation
ASIS	anterior superior iliac spine
ATFL	anterior talofibular ligament
ATLS	Advanced Trauma Life Support
AUSCAN	Australian–Canadian Hand Osteoarthritis Index
AVN	avascular necrosis
AVPU	aware, verbally responsive, pain responsive, and unresponsive
BAPRAS	British Association of Plastic, Reconstructive and Aesthetic Surgeons
BASICS	British Association for Immediate Care
BCIS	bone cement implantation syndrome
BCP	bicalcium phosphate
BMD	bone mineral density
BMI	body mass index
BMP	bone morphogenetic protein
BOA	British Orthopaedic Association
BOAST	BOA Standards for Trauma
BSA	body surface area
BSR	British Society for Rheumatology
BUN	blood urea nitrogen
BVM	bag–valve–mask
CaSR	calcium-sensing receptor
C-A-T™	Combat Application Tourniquet
CC	cartilage calcification
CCP	cyclic citrullinated peptide
CDH	congenital dislocation of the hip
CDR	cervical disc replacement
4CF	four-corner fusion
CIMT	constraint-induced movement therapy
CKD-MBD	chronic kidney disease mineral bone disorder
CMAP	compound muscle action potential

CMC	carpometacarpal
CMI	cell-mediated immunity
CNS	central nervous system
COC	ceramic on ceramic (THA bearing)
COMP	cartilage oligomeric matrix protein
COP	ceramic on polyethylene (THA bearing)
CORA	centre of rotation of angulation
COX-2	cyclooxygenase-2
CPM	continuous passive motion
CPPD	calcium pyrophosphate dihydrate
CR	cruciate retaining
CRP	C-reactive protein
CRPS	complex regional pain syndrome
CSF	cerebrospinal fluid
CT	computed tomography
CTX	serum type I collagen C-terminal cross-linking telopeptide
CVP	central venous pressure
DDD	degenerative disc disease
DDH	developmental dysplasia of the hip
DIC	disseminated intravascular coagulation
DIP(J)	distal interphalangeal (joint)
DISH	diffuse idiopathic skeletal hyperostosis
DISI	dorsal intercalated segment instability
DLC	*discoligamentous complex*
DLIF	direct lateral interbody fusion
DMARDs	disease-modifying antirheumatic drugs
DMD	Duchenne muscular dystrophy
DNA	deoxyribonucleic acid
DRUJ	distal radioulnar joint
DTH	delayed type hypersensitivity
DVT	deep vein thrombosis
DXA	dual-energy X-ray absorptiometry
ECRB	extensor carpi radialis brevis
ECRL	extensor carpi radialis longus
ECU	extensor carpi ulnaris
EDF	elongation–derotation–flexion
EEG	electroencephalography
eFAST	extended focused assessment sonography in trauma
eGFR	estimated glomerular filtration rate
EMG	electromyography
EMS	emergency medical service
EMT	emergency medical technician
ENL	erythema nodosum leprosum
ENT	ear, nose and throat
EPL	extensor pollicis longus
ESR	erythrocyte sedimentation rate
ETA	estimated time of arrival
$EtCO_2$	end-tidal carbon dioxide
EULAR	European League Against Rheumatism
FAB	foot abduction brace
FABER	Flexion, ABduction, and External Rotation test
FABS	flexion, abduction, supination
FAI	femoroacetabular impingement
FAST	focused assessment sonography in trauma
FBC	full blood count
FDP	flexor digitorum profundus
FDS	flexor digitorum superficialis
FFF-STA	Flat foot associated with a short tendo Achilles
FFO	functional foot orthoses
FGF	fibroblast growth factor
FGFR	fibroblast growth receptor
FHH	familial hypocalciuric hypercalcaemia
FHON	femoral head osteonecrosis
FISH	fluorescence in situ hybridization
FLS	Fracture Liaison Services
fMRI	functional magnetic resonance imaging
FMS	fibromyalgia syndrome
FNCLCC	Federation Nationale des Centres de Lutte Contre le Cancer
FPB	flexor pollicis brevis
FPE	fatal pulmonary embolism
FPL	flexor pollicis longus
GABA	gamma-aminobutryic acid
GAGs	glycosaminoglycans
GCS	Glasgow Coma Scale
GCT	giant cell tumour
GCTTS	giant cell tumour of tendon sheath
GMFCS	gross motor function classification system
GPI	general paralysis of the insane
GGT	gamma-glutamyl transferase
GH	*growth hormone*
GRF	ground reaction force
HA	hydroxyapatite
HEMS	helicopter emergency medical service
HHR	humeral head replacement
HIE	hypoxic–ischaemic encephalopathy
HIV	human immunodeficiency virus
HLA	human leucocyte antigen
HMSN	hereditary motor and sensory neuropathy
HNPP	hereditary neuropathy with liability to pressure palsies
HO	heterotopic ossification
HOOS	Hip Dysfunction and Osteoarthritis Outcome Score
HR	hip resurfacing
HRT	hormone replacement therapy
IASP	International Association for the Study of Pain
ICF	International Classification of Functioning, Disability and Health
ICP	intracerebral pressure
ICS	intercostal space
ICU	intensive care unit

IDH isocitrate dehydrogenase
IFSSH International Federation of Societies for Surgery of the Hand
IGRA interferon-gamma release assay
IL interleukin
IM intramuscular
IMRT intensity-modulated radiotherapy
INR international normalized ratio
IP(J) interphalangeal joint
IRIS immune reconstitution inflammatory syndrome
IRMER Ionising Radiation Medical Exposure Regulations
ISS injury severity score
ITB intrathecal baclofen
IV intervertebral *and* intravenous
IVF *in vitro* fertilization
IVH intraventricular haemorrhage
JIA juvenile idiopathic arthritis
JOAMEQ Japanese Orthopaedic Association Cervical Myelopathy Evaluation Questionnaire
KAFO knee–ankle–foot orthosis
KOOS Knee Dysfunction and Osteoarthritis Outcome Score
LBP lower back pain
LC lateral compression
LCH Langerhans cell histiocytosis
LCL lateral collateral ligament
LCPD Legg–Calvé–Perthes disease
LDH lactate dehydrogenase
LHB long head of biceps
LLD leg length discrepancy
LMA laryngeal mask airway
LMN lower motor neuron
LMWH low molecular weight heparin
MAP mean arterial pressure
MARS metal artifact reduction sequences (MRI)
MB multibacillary
MCL medial collateral ligament
MCP(J) metacarpophalangeal (joint)
M-CSF macrophage colony-stimulating factor
MCV mean corpuscular volume
MDM2 murine double minute-2
MED multiple epiphyseal dysplasia
MEN multiple endocrine neoplasia
MGUS monoclonal gammopathy of undetermined significance
MHC major histocompatibility complex
MIC minimal inhibitory concentration
MIPO minimally invasive percutaneous osteosynthesis
MND motor neuron disease
MO multiple osteochondromas
MODS multiple organ failure or dysfunction syndrome
MOM metal on metal (THA bearing)
MOP metal on polyethylene (THA bearing)
MP migration percentage
MPFL medial patellofemoral ligament
MPM mortality prediction model
MPNST malignant peripheral nerve sheath tumour
MPS mucopolysaccharidoses
MRC Medical Research Council
MRA magnetic resonance arthrography or angiography
MRI magnetic resonance imaging
MRSA methicillin-resistant *Staphylococcus aureus*
MSSA methicillin-sensitive *Staphylococcus aureus*
MTC Major Trauma Centre
MTP(J) metatarsophalangeal (joint)
NARU National Ambulance Resilience Unit
NCIN *National Cancer Intelligence Network*
NCTH non-compressible torso haemorrhage
NCV nerve conduction velocity
NDI Neck Disability Index
NF neurofibromatosis
NIBP non-invasive blood pressure
NICE National Institute for Health and Care Excellence
NOF non-ossifying fibroma
NP nasopharyngeal
NPS Nail–patella syndrome
NSAIDs non-steroidal anti-inflammatory drugs
OA osteoarthritis
OCD osteochondritis dissecans
OFD osteofibrous dysplasia
OI osteogenesis imperfecta
OMT Oberg, Manske and Tonkin (classification)
ONJ osteonecrosis of the jaw
OP oropharyngeal
OPG osteoprotegerin
OPLL ossification of the posterior longitudinal ligament
P1NP serum type I collagen extension propeptide
PA posteroanterior
PACS Picture Archiving and Communication System
PAFC pulmonary artery flotation catheterization
PAO periacetabular osteotomy
PAOP pulmonary artery occlusion pressure
PB paucibacillary
PCA patient-controlled analgesia
PCL posterior cruciate ligament
PCR polymerase chain reaction
PD proton density
PDB Paget's disease of bone

PE pulmonary embolism
PEA pulseless electrical activity
PEEP positive end-expiratory pressure
PET positron emission tomography
PH Pavlik harness
PHEM pre-hospital emergency medicine
Pi inorganic phosphate
PIP(J) proximal interphalangeal (joint)
PJI periprosthetic infection
PL posterolateral
PLC posterior ligamentous complex *and* posterolateral corner
PLL posterior longitudinal ligament
PLRI posterolateral rotatory instability
PM posteromedial
PMMA polymethylmethacrylate
PNS peripheral nervous system
PPE personal protective equipment
PPS post-polio syndrome
pQCT peripheral quantitative computer tomography
PRC proximal row carpectomy
PRICE protection, rest, ice, compression and elevation
PRICER protection, rest, ice, compression, elevation and rehabilitation
PRP platelet rich plasma
PS posterior stabilized
PSA prostate-specific antigen
PsA psoriatic arthritis
PTH parathyroid hormone
PTHrP parathyroid hormone-related peptide
PTS post-thrombotic syndrome
PVL periventricular leucomalacia
PVNS pigmented villonodular synovitis
QCT quantitative computed tomography
QoL quality of life
QUS quantitative ultrasonometry
RA radiographic absorptiometry *and* rheumatoid arthritis
RANKL receptor activator of nuclear factor-$\acute{\kappa}\beta$ ligand
REBOA resuscitative endovascular balloon occlusion of the aorta
RF rheumatoid factor
RGO reciprocating gait orthoses
RICE rest, ice, compression and elevation
RNA ribonucleic acid
RR reversal reaction
RSD reflex sympathetic dystrophy
RSI rapid sequence induction
RTC road traffic crash
SAC space available for spinal cord
SACE serum angiotensin converting enzyme
SAMU Services de l'Aide Medical Urgente
SAPHO synovitis, acne, pustulosis, hyperostosis and osteitis
SAPS simplified acute physiology score
SAS subaxial subluxation
SBC simple bone cyst
SCFE slipped capital femoral epiphysis
SCI spinal cord injury
SCIWORA spinal cord injury without obvious radiographic abnormality
SCM sternocleidomastoid muscle
SDD selective digestive tract decontamination
SDR selective dorsal rhizotomy
SE spin echo
SED spondyloepiphyseal dysplasia
SEMLS single event multi-level surgery
SERM selective oestrogen receptor modulator
SIJ sacroiliac joint
SIRS systemic inflammatory response
SLAP superior labrum, anterior and posterior (tear)
SLE systemic lupus erythematosus
SLIC Subaxial Cervical Spine Injury Classification
SMR standardized mortality ratio
SMUR Services Mobile d'Urgence et de Reamination
SNAP sensory nerve action potential
SNPs single nucleotide polymorphisms
SOFA sequential organ failure assessment
SONK 'spontaneous' osteonecrosis of the knee
SOP standard operating procedure
SPA spondyloarthropathy
SpA spondyloarthritis
SPECT single photon emission computed tomography
SPORT Spine Patient Outcomes Research Trial
STIR short-tau inversion recovery
STT scaphoid–trapezium–trapezoid arthritis *and* soft-tissue tumour
SUA serum uric acid
SUFE slipped upper femoral epiphysis
TAR thrombocytopaenia with absent radius syndrome
TARN Trauma Audit and Research Network
TB tuberculosis
TBI total body involvement
TBS Trabecular Bone Score
^{99m}Tc-HDP technetium(^{99m}Tc)-labelled hydroxymethylene diphosphonate
^{99m}Tc-MDP technetium(^{99m}Tc)-labelled methyl diphosphonate
TDR total disc replacement
TE time to echo
TFCC triangular fibrocartilage complex
THA total hip arthroplasty
TIP terminal interphalangeal (joint)

TISS therapeutic intervention scoring system
TKR total knee replacement
TLIF transforaminal lumbar interbody fusion
TMT tarsometatarsal
TNF tumour necrosis factor
TNM tumour–node–metastasis
TOE transoesophageal echocardiogram
TSF Taylor spatial frame
TSH thyroid-stimulating hormone
TSR total shoulder replacement
TU Trauma Unit
UCP unilateral cerebral palsy
UFD unifacet dislocation
UHMWPE ultra-high molecular weight polyethylene
UICC Union for International Cancer Control
ULT urate-lowering therapy
UMN upper motor neuron
UPS undifferentiated pleomorphic sarcoma
US ultrasound
VAC vacuum-assisted closure
VACTERLS refers to the systems involved and the defects identified: *v*ertebral, *a*nal, *c*ardiac, *t*racheal, *e*sophageal, *r*enal, *l*imb and *s*ingle umbilical artery
VCR vertebral column resection
VCT voluntary counselling and testing
VFA Vertebral Fracture Assessment
VISI volar intercalated segment instability
VMO vastus medialis oblique
VP ventriculoperitoneal
VQ ventilation–perfusion
VS vertical shear *and* vertical subluxation
VTE venous thromboembolism
WALANT wide awake local anaesthetic no tourniquet
WBC white blood cell
WHO World Health Organization
XLH sex-linked hypophosphataemic rickets

Taylor & Francis
Taylor & Francis Group
http://taylorandfrancis.com

Section 1

General Orthopaedics

Taylor & Francis
Taylor & Francis Group
http://taylorandfrancis.com

Diagnosis in orthopaedics

1

Louis Solomon & Charles Wakeley

Orthopaedics is concerned with bones, joints, muscles, tendons and nerves – the skeletal system and all that makes it move. Conditions that affect these structures fall into seven easily remembered pairs:

1 Congenital and developmental abnormalities
2 Infection and inflammation
3 Arthritis and rheumatic disorders
4 Metabolic and endocrine disorders
5 Tumours and lesions that mimic them
6 Neurological disorders and muscle weakness
7 Injury and mechanical derangement

Diagnosis in orthopaedics, as in all of medicine, is the identification of disease. It begins from the very first encounter with the patient and is gradually modified and fine-tuned until we have a picture, not only of a pathological process but also of the functional loss and the disability that goes with it. Understanding evolves from the systematic gathering of information from the history, the physical examination, tissue and organ imaging and special investigations. Systematic, but never mechanical; behind the enquiring mind there should also be what D. H. Lawrence has called 'the intelligent heart'. It must never be forgotten that the patient has a unique personality, a job and hobbies, a family and a home; all have a bearing upon, and are in turn affected by, the disorder and its treatment.

HISTORY

'Taking a history' is a misnomer. The patient tells a story; it is we the listeners who construct a history. The story may be maddeningly disorganized; the history has to be systematic. Carefully and patiently compiled, it can be every bit as informative as examination or laboratory tests.

As we record it, certain key words and phrases will inevitably stand out: injury, pain, stiffness, swelling, deformity, instability, weakness, altered sensibility and loss of function or inability to do certain things that were easily accomplished before.

Each symptom is pursued for more detail: we need to know when it began, whether suddenly or gradually, spontaneously or after some specific event; how it has changed or progressed; what makes it worse; what makes it better.

While listening, we consider whether the story fits some pattern that we recognize, for we are already thinking of a diagnosis. Every piece of information should be thought of as part of a larger picture which gradually unfolds in our understanding. The surgeon-philosopher Wilfred Trotter (1870–1939) put it well: 'Disease reveals itself in casual parentheses.'

Symptoms

Pain

Pain is the most common symptom in orthopaedics. It is usually described in metaphors that range from inexpressively bland to unbelievably bizarre – descriptions that tell us more about the patient's state of mind than about the physical disorder. Yet there are clearly differences between the throbbing pain of an abscess and the aching pain of chronic arthritis, between the 'burning pain' of neuralgia and the 'stabbing pain' of a ruptured tendon.

Severity is even more subjective. High and low pain thresholds undoubtedly exist, but pain is as bad as it feels to the patient, and any system of 'pain grading' must take this into account. The main value of estimating severity is in assessing the progress of the disorder or the response to treatment. The commonest method is to invite the patient to mark the severity on an analogue scale of 1–10, with 1 being mild and easily ignored, and 10 being totally unbearable. The problem about this type of grading is that patients who have never experienced very severe pain simply do not know what 8 or 9 or

10 would feel like. The following is suggested as a simpler system:

Grade I (mild) Pain that can easily be ignored
Grade II (moderate) Pain that cannot be ignored, interferes with function and needs attention or treatment from time to time
Grade III (severe) Pain that is present most of the time, demanding constant attention or treatment
Grade IV (excruciating) Totally incapacitating pain

Identifying the site of pain may be equally vague. Yet its precise location is important, and in orthopaedics it is useful to ask the patient to point to – rather than to say – where it hurts. Even then, do not assume that the site of pain is necessarily the site of pathology; 'referred' pain and 'autonomic' pain can be very deceptive.

Referred pain Pain arising in or near the skin is usually localized accurately. Pain arising in deep structures is more diffuse and is sometimes of unexpected distribution; thus, hip disease may manifest with pain in the knee (so might an obturator hernia). This is not because sensory nerves connect the two sites; it is due to inability of the cerebral cortex to differentiate clearly between sensory messages from separate but embryologically related sites. A common example is 'sciatica' – pain at various points in the buttock, thigh and leg, supposedly following the course of the sciatic nerve. Such pain is not necessarily due to pressure on the sciatic nerve or the lumbar nerve roots; it may be 'referred' from any one of a number of structures in the lumbar spine, the pelvis and the posterior capsule of the hip joint. See Figure 1.1.

Autonomic pain We are so accustomed to matching pain with some discrete anatomical structure and its known sensory nerve supply that we are apt to dismiss any pain that does not fit the usual pattern as 'atypical' or 'inappropriate' (i.e. psychologically determined).

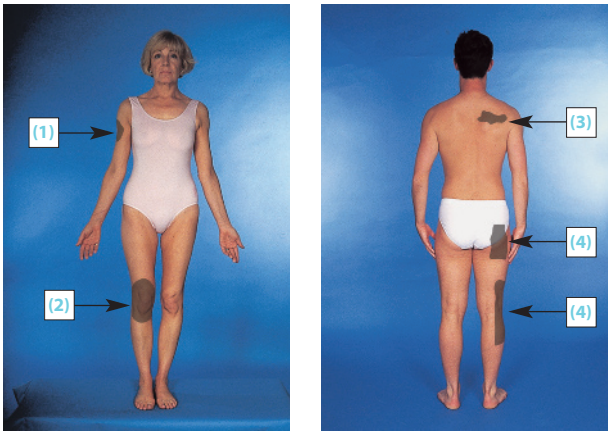


Figure 1.1 Referred pain Common sites of referred pain: (1) from the shoulder; (2) from the hip; (3) from the neck; (4) from the lumbar spine.

But pain can also affect the autonomic nerves that accompany the peripheral blood vessels and this is much more vague, more widespread and often associated with vasomotor and trophic changes. It is poorly understood, often doubted, but nonetheless real.

Stiffness

Stiffness may be generalized (typically in systemic disorders such as rheumatoid arthritis and ankylosing spondylitis) or localized to a particular joint. Patients often have difficulty in distinguishing localized stiffness from painful movement; limitation of movement should never be assumed until verified by examination.

Ask when it occurs: regular early morning stiffness of many joints is one of the cardinal symptoms of rheumatoid arthritis, whereas transient stiffness of one or two joints after periods of inactivity is typical of osteoarthritis.

Locking 'Locking' is the term applied to the sudden inability to complete a particular movement. It suggests a mechanical block – for example, due to a loose body or a torn meniscus becoming trapped between the articular surfaces of the knee. Unfortunately, patients tend to use the term for any painful limitation of movement; much more reliable is a history of 'unlocking', when the offending body slips out of the way.

Swelling

Swelling may be in the soft tissues, the joint or the bone; to the patient they are all the same. It is important to establish whether it followed an injury, whether it appeared rapidly (think of a haematoma or a haemarthrosis) or slowly (due to inflammation, a joint effusion, infection or a tumour), whether it is painful (suggestive of acute inflammation, infection or a tumour), whether it is constant or comes and goes, and whether it is increasing in size.

Deformity

The common deformities are described by patients in terms such as round shoulders, spinal curvature, knock knees, bow legs, pigeon toes and flat feet. Deformity of a single bone or joint is less easily described and the patient may simply declare that the limb is 'crooked'.

Some 'deformities' are merely variations of the normal (e.g. short stature or wide hips); others disappear spontaneously with growth (e.g. flat feet or bandy legs in an infant). However, if the deformity is progressive, or if it affects only one side of the body while the opposite joint or limb is normal, it may be serious (Figure 1.2).

Weakness

Generalized weakness is a feature of all chronic illness, and any prolonged joint dysfunction will inevitably

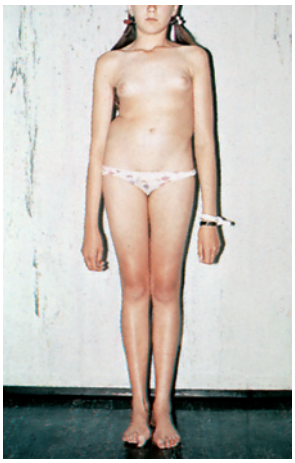



Figure 1.2 Deformity This young girl complained of a prominent right hip; the real deformity was scoliosis.

lead to weakness of the associated muscles. However, pure muscular weakness – especially if it is confined to one limb or to a single muscle group – is more specific and suggests some neurological or muscle disorder. Patients sometimes say that the limb is 'dead' when it is actually weak, and this can be a source of confusion. Questions should be framed to discover precisely which movements are affected, for this may give important clues, if not to the exact diagnosis at least to the site of the lesion.

Instability

The patient may complain that the joint 'gives way' or 'jumps out of place'. If this happens repeatedly, it suggests abnormal joint laxity, capsular or ligamentous deficiency, or some type of internal derangement such as a torn meniscus or a loose body in the joint. If there is a history of injury, its precise nature is important.

Change in sensibility

Tingling or numbness signifies interference with nerve function – pressure from a neighbouring structure (e.g. a prolapsed intervertebral disc), local ischaemia (e.g. nerve entrapment in a fibro-osseous tunnel) or a peripheral neuropathy. It is important to establish its exact distribution; from this we can tell whether the fault lies in a peripheral nerve or in a nerve root. We should also ask what makes it worse or better; a change in posture might be the trigger, thus focusing attention on a particular site.

Loss of function

Functional disability is more than the sum of individual symptoms and its expression depends upon the needs of that particular patient. The patient may say, 'I can't stand for long' rather than 'I have backache'; or 'I can't put my socks on' rather than 'My hip is stiff.' Moreover, what to one patient is merely inconvenient may, to another, be incapacitating. Thus a lawyer or a teacher may readily tolerate a stiff knee provided it is painless, but to a plumber or a parson the same disorder might spell economic or spiritual disaster. One question should elicit the important information: 'What can't you do now that you used to be able to do?'

Past history

Patients often forget to mention previous illnesses or accidents, or they may simply not appreciate their relevance to the present complaint. They should be asked specifically about childhood disorders, periods of incapacity and old injuries. A 'twisted ankle' many years ago may be the clue to the onset of osteoarthritis in what is otherwise an unusual site for this condition. Gastrointestinal disease, which in the patient's mind has nothing to do with bones, may be important in the later development of ankylosing spondylitis or osteoporosis. Similarly, certain rheumatic disorders may be suggested by a history of conjunctivitis, iritis, psoriasis or urogenital disease. Metastatic bone disease may erupt many years after a mastectomy for breast cancer. Patients should also be asked about previous medication: many drugs, and especially corticosteroids, have long-term effects on bone. Alcohol and drug abuse are important, and we must not be afraid to ask about them.

Family history

Patients often wonder (and worry) about inheriting a disease or passing it on to their children. To the doctor, information about musculoskeletal disorders in the patient's family may help with both diagnosis and counselling. When dealing with a suspected case of bone or joint infection, ask about communicable diseases, such as tuberculosis or sexually transmitted disease, in other members of the family.

Social background

No history is complete without enquiry about the patient's background. There are the obvious things such as the level of care and nutrition in children; dietary constraints which may cause specific deficiencies; and, in certain cases, questions about smoking habits, alcohol consumption and drug abuse, all of which call for a special degree of tact and non-judgemental enquiry.

Find out details about the patient's work practices, travel and recreation: could the disorder be due to

a particular repetitive activity in the home, at work or on the sports field? Is the patient subject to any unusual occupational strain? Has he or she travelled to another country where tuberculosis is common?

Finally, it is important to assess the patient's home circumstances and the level of support by family and friends. This will help to answer the question: 'What has the patient lost and what is he or she hoping to regain?'

EXAMINATION

In *A Case of Identity*, Sherlock Holmes has the following conversation with Dr Watson.

Watson: You appeared to read a good deal upon [your client] which was quite invisible to me.

Holmes: Not invisible but unnoticed, Watson.

Some disorders can be diagnosed at a glance: who would mistake the facial appearance of acromegaly or the hand deformities of rheumatoid arthritis for anything else? Nevertheless, even in these cases systematic examination is rewarding: it provides information about the patient's particular disability, as distinct from the clinicopathological diagnosis; it keeps reinforcing good habits; and, never to be forgotten, it lets the patient know that he or she has been thoroughly attended to.

The examination actually begins from the moment we set eyes on the patient. We observe his or her general appearance, posture and gait. Can you spot any distinctive feature: Knock-knees? Spinal curvature? A short limb? A paralysed arm? Does he or she appear to be in pain? Do their movements look natural? Do they walk with a limp, or use a stick? A telltale gait may suggest a painful hip, an unstable knee or a foot-drop. The clues are endless and the game is played by everyone (qualified or lay) at each new encounter throughout life. In the clinical setting the assessment needs to be more focused.

When we proceed to the structured examination, the patient must be suitably undressed; no mere rolling up of a trouser leg is sufficient. If one limb is affected, both must be exposed so that they can be compared.

We examine the good limb (for comparison), then the bad. There is a great temptation to rush in with both hands – a temptation that must be resisted. Only by proceeding in a purposeful, orderly way can we avoid missing important signs.

Alan Apley, who developed and taught the system used here, shied away from using long words where short ones would do the job. (He also used to say, 'I'm neither an inspector nor a manipulator, and I am definitely not a palpator.') Thus the traditional clinical routine, inspection, palpation, manipulation, was replaced by *look*, *feel*, *move*. With time, his teaching has been extended and we now add *test*, to include the special manoeuvres we employ in assessing neurological integrity and complex functional attributes.

Look

Abnormalities are not always obvious at first sight. A systematic, step-by-step process helps to avoid mistakes.

Shape and posture The first things to catch one's attention are the shape and posture of the limb or the body or the entire person who is being examined. Is the patient unusually thin or obese? Does the overall posture look normal? Is the spine straight or unusually curved? Are the shoulders level? Are the limbs normally positioned? It is important to look for deformity in three planes, and always compare the affected part with the normal side. In many joint disorders and in most nerve lesions the limb assumes a characteristic posture. In spinal disorders the entire torso may be deformed. Now look more closely for swelling or wasting – one often enhances the appearance of the other! Or is there a definite lump?

Skin Careful attention is paid to the colour, quality and markings of the skin. Look for bruising, wounds and ulceration. Scars are an informative record of the past – surgical archaeology, so to speak (see Figure 1.3). Colour reflects vascular status or pigmentation – for example, the pallor of ischaemia, the blueness of cyanosis, the redness of inflammation, or the dusky purple of an old bruise. Abnormal creases, unless due to fibrosis, suggest underlying deformity which is not always obvious; tight, shiny skin with no creases is typical of oedema or trophic change.

General survey Attention is initially focused on the symptomatic or most obviously abnormal area, but we

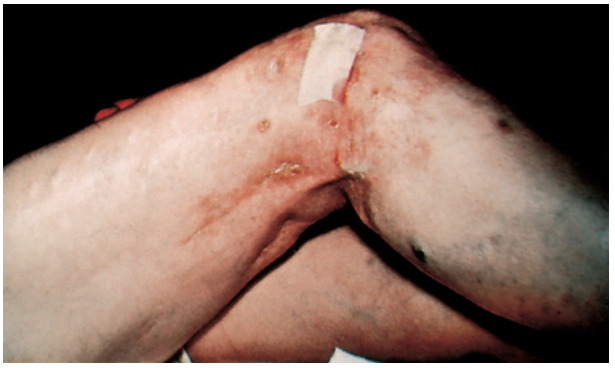


Figure 1.3 Look Scars often give clues to the previous history. The faded scar on this patient's thigh is an old operation wound – internal fixation of a femoral fracture. The other scars are due to postoperative infection; one of the sinuses is still draining.

must also look further afield. The patient complains of the joint that is hurting now, but we may see at a glance that several other joints are affected as well.

Feel

Feeling is exploring, not groping aimlessly. Know your anatomy and you will know where to feel for the landmarks; find the landmarks and you can construct a virtual anatomical picture in your mind's eye.

The skin Is it warm or cold; moist or dry; and is sensation normal?

The soft tissues Can you feel a lump; if so, what are its characteristics? Are the pulses normal?

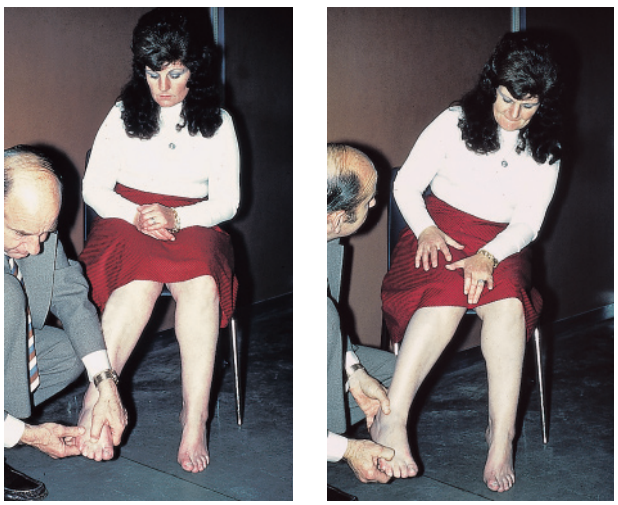
The bones and joints Are the outlines normal? Is the synovium thickened? Is there excessive joint fluid?

Tenderness Once you have a clear idea of the structural features in the affected area, feel gently for tenderness (Figure 1.4). Keep your eyes on the patient's face; a grimace will tell you as much as a grunt. Try to localize any tenderness to a particular structure; if you know precisely *where* the trouble is, you are halfway to knowing *what* it is.

Move

'Movement' covers several different activities: active movement, passive movement, abnormal or unstable movement, and provocative movement (see Figures 1.5 and 1.6).

Active movement Ask the patient to move without your assistance. This will give you an idea of the degree of mobility and whether it is painful or not. Active movement is also used to assess muscle power.



(a) (b)

Figure 1.4 Feeling for tenderness (a) The wrong way – there is no need to look at your fingers, you should know where they are. (b) It is much more informative to look at the patient's face!

Passive movement Here it is the examiner who moves the joint in each anatomical plane. Note whether there is any difference between the range of active and passive movement.

Range of movement is recorded in degrees, starting from zero which, by convention, is the neutral or anatomical position of the joint, and finishing where movement stops, due either to pain or to anatomical limitation. Describing the range of movement is often made to seem difficult. Words such as 'full', 'good', 'limited' and 'poor' are misleading. Always cite the range or span, from start to finish, in degrees. For example, 'knee flexion 0–140 degrees' means that the range of flexion is from zero (the knee absolutely straight) through an arc of 140 degrees (the leg making an acute angle with the thigh). Similarly, 'knee flexion 20–90 degrees' means that flexion begins at 20 degrees (i.e. the joint cannot extend fully) and continues only to 90 degrees.

For accuracy you can measure the range of movement with a goniometer, but with practice you will learn to estimate the angles by eye. Normal ranges of movement are shown in chapters dealing with individual joints. What is important is always to compare the symptomatic with the asymptomatic or normal side.

While testing movement, feel for crepitus. Joint crepitus is usually coarse and fairly diffuse; tenosynovial crepitus is fine and precisely localized to the affected tendon sheath.

Unstable movement This is movement which is inherently unphysiological. You may be able to shift or angulate a joint out of its normal plane of movement, thus demonstrating that the joint is unstable. Such abnormal movement may be obvious (e.g. a wobbly knee); often, though, you have to use special manoeuvres to pick up minor degrees of instability.

Provocative movement One of the most telling clues to diagnosis is reproducing the patient's symptoms by applying a specific, provocative movement. Shoulder pain due to impingement of the subacromial structures may be 'provoked' by moving the joint in a way that is calculated to produce such impingement; the patient recognizes the similarity between this pain and his or her daily symptoms. Likewise, a patient who has had a previous dislocation or subluxation can be vividly reminded of that event by stressing the joint in such a way that it again threatens to dislocate; indeed, merely starting the movement may be so distressing that the patient goes rigid with anxiety at the anticipated result – this is aptly called the *apprehension test.*

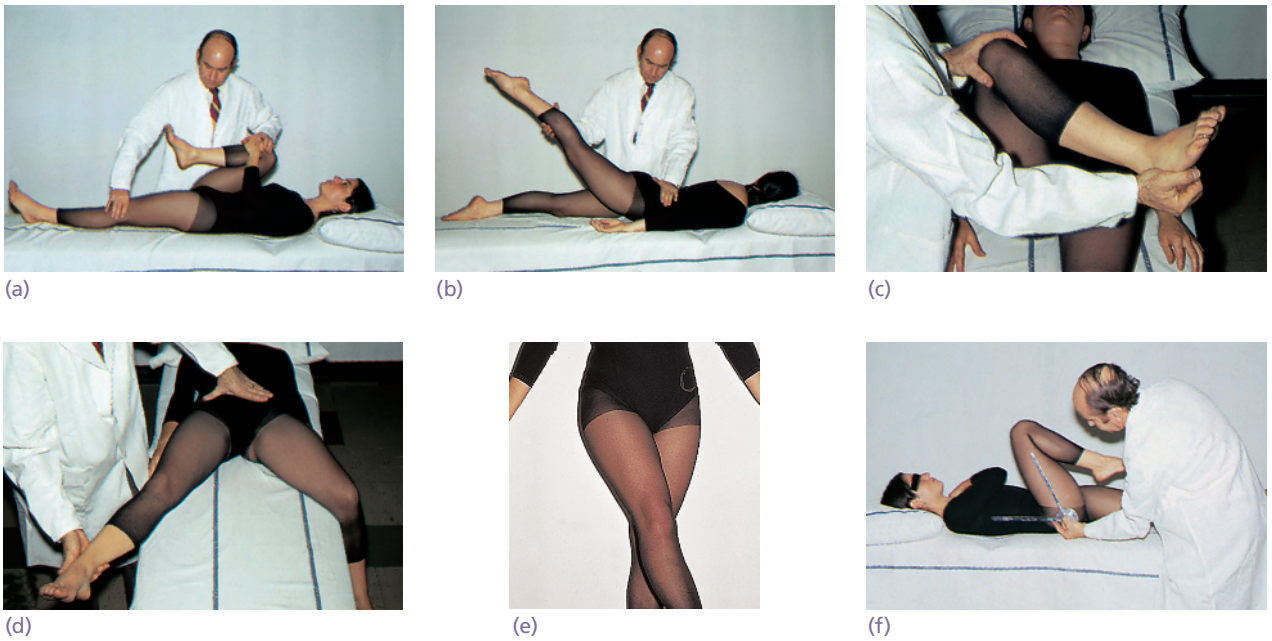


Figure 1.5 Testing for movement (a) Flexion, (b) extension, (c) rotation, (d) abduction, (e) adduction. The range of movement can be estimated by eye or measured accurately using a goniometer (f).

Test

The apprehension test referred to in the previous paragraph is one of several clinical tests that are used to elicit suspected abnormalities: some examples are *Thomas' test* for flexion deformity of the hip, *Trendelenburg's test* for instability of the hip, *McMurray's test* for a torn meniscus of the knee, *Lachman's test* for cruciate ligament instability and various tests for intra-articular fluid. These and others are described in the relevant chapters in Section 2. Tests for muscle tone, motor power, reflexes and various modes of sensibility are part and parcel of neurological examination, which is discussed later in this chapter.

Caveat

We recognize that the sequence set out here may sometimes have to be modified. We may need to 'move' before we 'look': an early scoliotic deformity of the spine often becomes apparent only when the patient bends forwards. The sequence may also have to be altered because a patient is in severe pain or disabled: you would not try to move a limb at all in someone with a suspected fracture when an X-ray can provide the answer. When examining a child, you may have to take your chances with look or feel or move whenever you can!

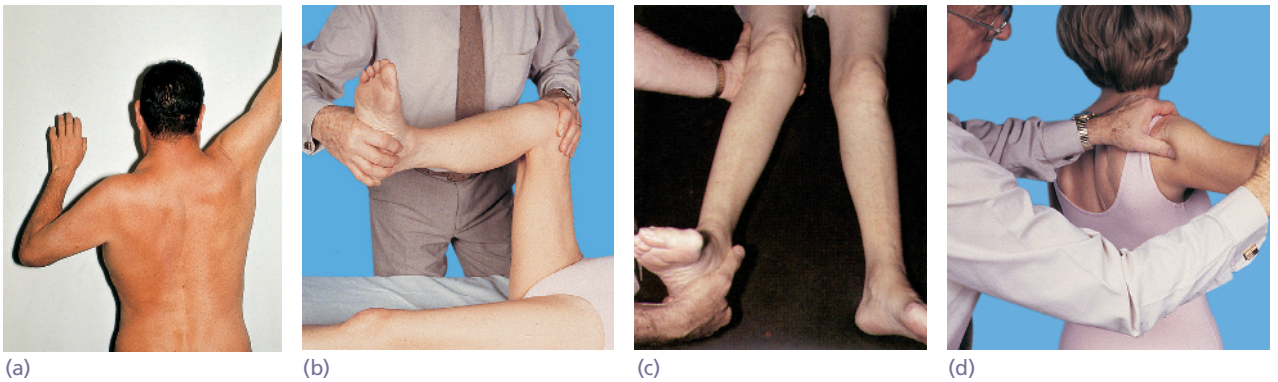


Figure 1.6 Move (a) Active movement – the patient moves the joint. The right shoulder is normal; the left has restricted active movement. (b) Passive movement – the examiner moves the joint. (c) Unstable movement – the joint can be moved across the normal planes of action, in this case demonstrating valgus instability of the right knee. (d) Provocative movement – the examiner moves (or manipulates) the joint so as to provoke the symptoms of impending pain or dislocation. Here he is reproducing the position in which an unstable shoulder is likely to dislocate.

Terminology

Colloquial terms such as front, back, upper, lower, inner aspect, outer aspect, bow legs, knock knees have the advantage of familiarity but are not applicable to every situation. Universally acceptable anatomical definitions are therefore necessary in describing physical attributes.

Bodily surfaces, planes and positions are always described in relation to the **anatomical position** – as if the person were standing erect, facing the viewer, legs together with the knees pointing directly forwards, and arms held by the sides with the palms facing forwards.

The principal **planes** of the body (Figure 1.7) are named **sagittal, coronal and transverse**; they define the direction across which the body (or body part) is viewed in any description. **Sagittal planes**, parallel to each other, pass vertically through the body from front to back; the **midsagittal** or **median plane** divides the body into right and left halves. **Coronal planes** are also orientated vertically, corresponding to a frontal view, at right angles to the sagittal planes; **transverse planes** pass horizontally across the body.

Anterior signifies the frontal aspect and **posterior** the rear aspect of the body or a body part. The terms **ventral** and **dorsal** are also used for the front and the back respectively. Note, though, that the use of these terms is somewhat confusing when it comes to the foot: here the upper surface is called the **dorsum** and the sole is called the **plantar surface**.

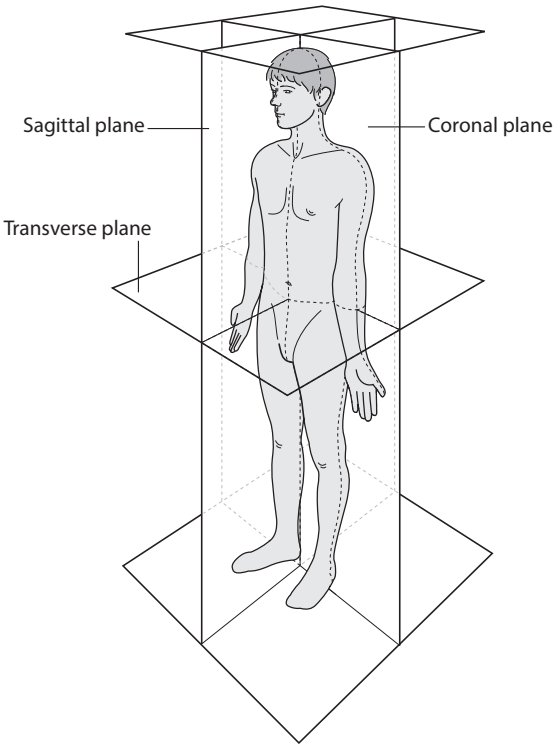


FIgure 1.7 Planes The principal planes of the body, as viewed in the anatomical position: sagittal, coronal and transverse.

Medial means facing towards the median plane or midline of the body, and **lateral** away from the median plane. These terms are usually applied to a limb, the clavicle or one half of the pelvis. Thus the inner aspect of the thigh lies on the medial side of the limb and the outer part of the thigh lies on the lateral side. We could also say that the little finger lies on the medial or **ulnar side** of the hand and the thumb on the lateral or **radial side** of the hand.

Proximal and **distal** are used mainly for parts of the limbs, meaning respectively the upper end and the lower end as they appear in the anatomical position. Thus the knee joint is formed by the distal end of the femur and the proximal end of the tibia.

Axial alignment describes the longitudinal arrangement of adjacent limb segments or parts of a single bone. The knees and elbows, for example, are normally angulated slightly outwards (**valgus**) while the opposite – 'bow legs' – is more correctly described as **varus** (see 'Physical variations and deformities' later in this chapter). Angulation in the middle of a long bone would always be regarded as abnormal.

Rotational alignment refers to the tortile arrangement of segments of a long bone (or an entire limb) around a single longitudinal axis. For example, in the anatomical position the patellae face forwards while the feet are turned slightly outwards; a marked difference in rotational alignment of the two legs is abnormal.

Flexion and extension are joint movements in the sagittal plane, most easily imagined in hinge joints like the knee, elbow and the joints of the fingers and toes. In elbows, knees, wrists and fingers, flexion means bending the joint and extension means straightening it. In shoulders and hips, flexion is movement in an anterior direction and extension is movement posteriorwards. In the ankle, flexion is also called **plantarflexion** (pointing the foot downwards) and extension is called **dorsiflexion** (drawing the foot upwards). Thumb movements are the most complicated and are described in Chapter 16.

Abduction and adduction are movements in the coronal plane, away from or towards the median plane. Not quite for the fingers and toes, though: here abduction and adduction mean away from and towards the longitudinal midline of the hand or foot!

Lateral rotation and medial rotation are twisting movements, outwards and inwards, around a longitudinal axis.

Pronation and supination are also rotatory movements, but the terms are applied only to movements of the forearm and the foot.

Circumduction is a composite movement made up of a rhythmic sequence of all the other movements. It is possible only for ball-and-socket joints such as the hip and shoulder.

Specialized movements such as opposition of the thumb, lateral flexion and rotation of the spine, and inversion or eversion of the foot, will be described in the relevant chapters.

NEUROLOGICAL EXAMINATION

If the symptoms include weakness or incoordination or a change in sensibility, or if they point to any disorder of the neck or back, a complete neurological examination of the related part is mandatory. Once again we follow a systematic routine, first looking at the general appearance, then assessing motor function (muscle tone, power and reflexes) and finally testing for sensory function (both skin sensibility and deep sensibility) (see Table 1.1 and Figure 1.8).

Table 1.1 Nerve root supply and actions of main muscle groups

Muscles/Muscle action	Nerve root supply
Sternomastoids	Spinal accessory C2, 3, 4
Trapezius	Spinal accessory C3, 4
Diaphragm	C3, 4, 5
Deltoid	C5, 6
Supra- and infraspinatus	C5, 6
Serratus anterior	C5, 6, 7
Pectoralis major	C5, 6, 7, 8
Elbow flexion extension	C5, 6 C7
Supination	C5, 6
Pronation	C6
Wrist extension flexion	C6, (7) C7, (8)
Finger extension flexion ab- and adduction	C7 C7, 8, T1 C8, T1
Hip flexion extension adduction abduction	L1, 2, 3 L5, S1 L2, 3, 4 L4, 5, S1
Knee extension flexion	L(2), 3, 4 L5, S1
Ankle dorsiflexion plantarflexion inversion eversion	L4, 5 S1, 2 L4, 5 L5, S1
Toe extension flexion abduction	L5 S1 S1, 2

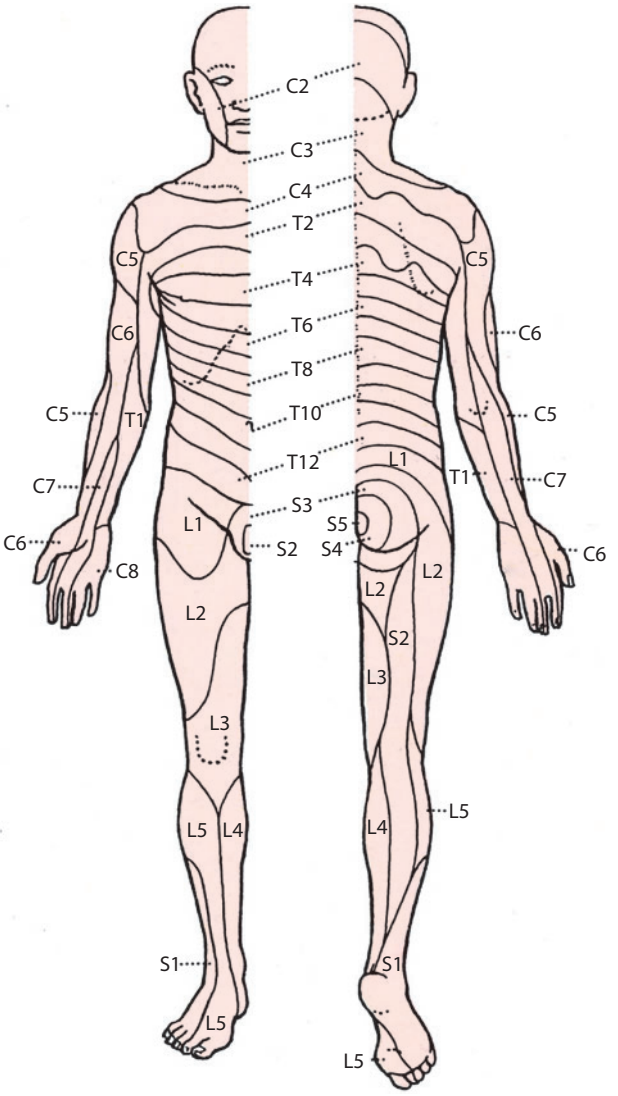


FIgure 1.8 Examination Dermatomes supplied by the spinal nerve roots.

Appearance

Some neurological disorders result in postures that are so characteristic as to be diagnostic at a glance: the claw hand of an ulnar nerve lesion; 'drop wrist' following radial nerve palsy (Figure 1.9); or the 'waiter's tip' deformity of the arm in brachial plexus injury. Usually, however, it is when the patient moves that we can best appreciate the type and extent of motor disorder: the dangling arm following a brachial plexus injury; the flail lower limb of poliomyelitis; the symmetrical paralysis of spinal cord lesions; the characteristic drop-foot gait following sciatic or peroneal nerve damage; and the jerky, 'spastic' movements of cerebral palsy.

Concentrating on the affected part, we look for trophic changes that signify loss of sensibility: the smooth, hairless skin that seems to be stretched too tight; atrophy of the fingertips and the nails; scars

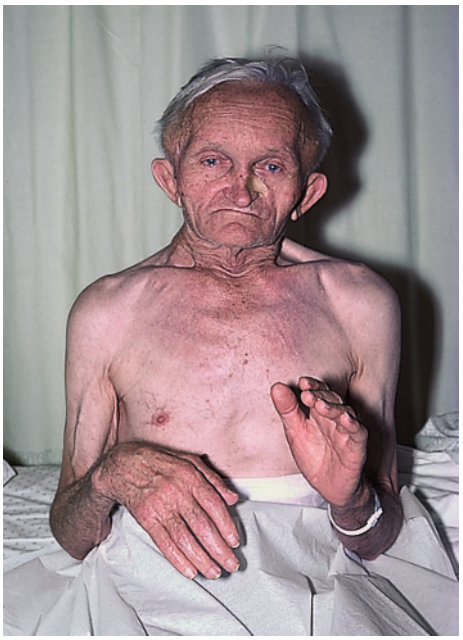


FIgure 1.9 Posture Posture is often diagnostic. This patient's 'drop wrist' – typical of a radial nerve palsy – is due to carcinomatous infiltration of the supraclavicular lymph nodes on the right.

that tell of accidental burns; and ulcers that refuse to heal. Muscle wasting is important: if localized and asymmetrical, it may suggest dysfunction of a specific motor nerve.

Muscle tone

Tone in individual muscle groups is tested by moving the nearby joint to stretch the muscle. Increased tone (spasticity) is characteristic of upper motor neuron disorders such as cerebral palsy and stroke. It must not be confused with rigidity (the 'lead-pipe' or 'cogwheel' effect) which is seen in Parkinson's disease. Decreased tone (flaccidity) is found in lower motor neuron lesions; for example, poliomyelitis. Muscle power is diminished in all three states; it is important to recognize that a 'spastic' muscle may still be weak.

Power

Motor function is tested by having the patient perform movements that are normally activated by specific nerves. We may learn even more about composite movements by asking the patient to perform specific tasks, such as holding a pen, gripping a rod, doing up a button or picking up a pin.

Testing for power is not as easy as it sounds; the difficulty is making ourselves understood. The simplest way is to place the limb in the 'test' position, then ask the patient to hold it there as firmly as possible and resist any attempt to change that position. The normal limb is examined first, then the affected limb, and the two are compared. Finer muscle actions, such as those of the thumb and fingers, may be reproduced by first demonstrating the movement yourself, then testing it in the unaffected limb, and then in the affected one.

Muscle power is usually graded on the Medical Research Council scale:

Grade 0	No movement
Grade 1	Only a flicker of movement
Grade 2	Movement with gravity eliminated
Grade 3	Movement against gravity
Grade 4	Movement against resistance
Grade 5	Normal power

It is important to recognize that muscle weakness may be due to muscle disease rather than nerve disease. In muscle disorders the weakness is usually more widespread and symmetrical, and sensation is normal.

Tendon reflexes

A deep tendon reflex is elicited by rapidly stretching the tendon near its insertion. A sharp tap with the tendon hammer does this well; but all too often this is performed with a flourish and with such force that the finer gradations of response are missed. It is better to employ a series of taps, starting with the most forceful and reducing the force with each successive tap until there is no response. Comparing the two sides in this way, we can pick up fine differences showing that a reflex is 'diminished' rather than 'absent'. In the upper limb we test biceps, triceps and brachioradialis; and in the lower limb the patellar and Achilles tendons.

The tendon reflexes are monosynaptic segmental reflexes; that is, the reflex pathway takes a 'short cut' through the spinal cord at the segmental level. Depression or absence of the reflex signifies interruption of the pathway at the posterior nerve root, the anterior horn cell, the motor nerve root or the peripheral nerve. It is a reliable pointer to the segmental level of dysfunction: thus, a depressed biceps jerk suggests pressure on the fifth or sixth cervical (C5 or C6) nerve roots while a depressed ankle jerk signifies a similar abnormality at the first sacral level (S1). An unusually brisk reflex, on the other hand, is characteristic of an upper motor neuron disorder (e.g. cerebral palsy, a stroke or injury to the spinal cord); the lower motor neuron is released from the normal central inhibition and there is an exaggerated response to tendon stimulation. This may manifest as ankle clonus: a sharp upward jerk on the foot (dorsiflexion) causes a repetitive, 'clonic' movement of the foot; similarly, a sharp downward push on the patella may elicit patellar clonus.

Superficial reflexes

The superficial reflexes are elicited by stroking the skin at various sites to produce a specific muscle contraction; the best known are the abdominal (T7–T12), cremasteric (L1, 2) and anal (S4, 5) reflexes. These are corticospinal (upper motor neuron) reflexes. Absence of the reflex indicates an upper motor neuron lesion (usually in the spinal cord) above that level.

The plantar reflex

Forceful stroking of the sole normally produces flexion of the toes (or no response at all). An extensor response (the big toe extends while the others remain in flexion) is characteristic of upper motor neuron disorders. This is the *Babinski sign* – a type of withdrawal reflex which is present in young infants and normally disappears after the age of 18 months.

Sensibility

Sensibility to touch and to pinprick may be increased (hyperaesthesia) or unpleasant (dysaesthesia) in certain irritative nerve lesions. More often, though, it is diminished (hypoaesthesia) or absent (anaesthesia), signifying pressure on or interruption of a peripheral nerve, a nerve root or the sensory pathways in the spinal cord. The area of sensory change can be mapped out on the skin and compared with the known segmental or dermatomal pattern of innervation. If the abnormality is well defined, it is an easy matter to establish the level of the lesion, even if the precise cause remains unknown.

Brisk percussion along the course of an injured nerve may elicit a tingling sensation in the distal distribution of the nerve (*Tinel's sign*). The point of hypersensitivity marks the site of abnormal nerve sprouting: if it progresses distally at successive visits, this signifies regeneration; if it remains unchanged, this suggests a local neuroma.

Tests for temperature recognition and two-point discrimination (the ability to recognize two touch-points a few millimetres apart) are also used in the assessment of peripheral nerve injuries.

Deep sensibility can be examined in several ways. In the vibration test a sounded tuning fork is placed over a peripheral bony point (e.g. the medial malleolus or the head of the ulna); the patient is asked if he or she can feel the vibrations and to say when they disappear. By comparing the two sides, differences can be noted. Position sense is tested by asking the patient to find certain points on the body with the eyes closed – for example, touching the tip of the nose with the forefinger. The sense of joint posture is tested by grasping the big toe and placing it in different positions of flexion and extension. The patient (whose eyes are closed) is asked to say whether it is 'up' or 'down'. Stereognosis, the ability to recognize shape and texture by feel alone, is tested by giving the patient (again with eyes closed) a variety of familiar objects to hold and asking him or her to name each object.

The pathways for deep sensibility run in the posterior columns of the spinal cord. Disturbances are therefore found in peripheral neuropathies and in spinal cord lesions such as posterior column injuries or tabes dorsalis. The sense of balance is also carried in the posterior columns. This can be tested by asking the patient to stand upright with his or her eyes closed; excessive body sway is abnormal (*Romberg's sign*).

Cortical and cerebellar function

A staggering gait may imply an unstable knee – or a disorder of the spinal cord or cerebellum. If there is no musculoskeletal abnormality to account for the sign, a full examination of the central nervous system will be necessary.

EXAMINING INFANTS AND CHILDREN

Paediatric practice requires special skills. You may have no first-hand account of the symptoms; a baby screaming with pain will tell you very little, and over-anxious parents will probably tell you too much. When examining the child, be flexible. If he or she is moving a particular joint, take your opportunity to examine movement then and there. You will learn much more by adopting methods of play than by applying a rigid system of examination. And leave any test for tenderness until last!

Infants and small children

The baby should be undressed, in a warm room, and placed on the examining couch. Look carefully for birthmarks, deformities and abnormal movements – or absence of movement. If there is no urgency or distress, take time to examine the head and neck, including facial features which may be characteristic of specific dysplastic syndromes. The back and limbs are then examined for abnormalities of position or shape.

Examining for joint movement can be difficult. Active movements can often be stimulated by gently stroking the limb. When testing for passive mobility, be careful to avoid frightening or hurting the child. In the neonate, and throughout the first two years of life, examination of the hips is mandatory, even if the child appears to be normal. This is to avoid missing the subtle signs of developmental dysplasia of the hips (DDH) at the early stage when treatment is most

Table 1.2 Normal developmental milestones

Age	Normal developmental milestone(s)
Newborn	Grasp reflex present Morrow reflex present
3–6 months	Holds head up unsupported
6–9 months	Able to sit up
9–12 months	Crawling Standing up
9–18 months	Walking
18–24 months	Running

effective. It is also important to assess the child's general development by testing for the normal milestones which are expected to appear during the first two years of life (Table 1.2).

Older children

Most children can be examined in the same way as adults, though with different emphasis on particular physical features. Posture and gait are very important; subtle deviations from the norm may herald the appearance of serious abnormalities such as scoliosis or neuromuscular disorders, while more obvious 'deformities' such as knock knees and bow legs may be no more than transient stages in normal development; similarly with mild degrees of 'flat feet' and 'pigeon toes'. More complex variations in posture and gait patterns, when the child sits and walks with the knees turned inwards (medially rotated) or outwards (laterally rotated) are usually due to anteversion or retroversion of the femoral necks, sometimes associated with compensatory rotational 'deformities' of the femora and tibiae. Seldom need anything be done about this; the condition usually improves as the child approaches puberty and only if the gait is very awkward would one consider performing corrective osteotomies of the femora.

PHYSICAL VARIATIONS AND DEFORMITIES

Joint laxity

Children's joints are much more mobile than those of most adults, allowing them to adopt postures that would be impossible for their parents. An unusual degree of joint mobility can also be attained by adults willing to submit to rigorous exercise and practice, as witness the performances of professional dancers and athletes, but in most cases, when the exercises stop, mobility gradually reverts to the normal range.

Persistent generalized joint hypermobility occurs in about 5% of the population and is inherited as a simple Mendelian dominant (Figure 1.10). Those affected

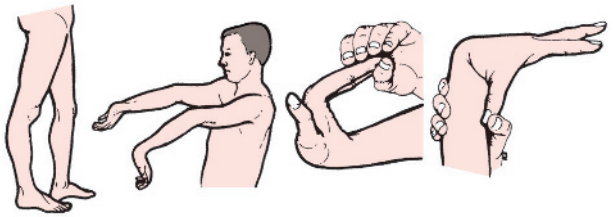


Figure 1.10 Tests for joint hypermobility
Hyperextension of knees and elbows; metacarpophalangeal joints extending to 90 degrees'; thumb able to touch forearm.

describe themselves as being 'double-jointed': they can hyperextend their metacarpophalangeal joints beyond a right angle, hyperextend their elbows and knees and bend over with knees straight to place their hands flat on the ground; some can even 'do the splits' or place their feet behind their neck!

It is doubtful whether these individuals should be considered 'abnormal'. However, epidemiological studies have shown that they do have a greater than usual tendency to recurrent dislocation (e.g. of the shoulder or patella). Some experience recurrent episodes of aching around the larger joints; however, there is no convincing evidence that hypermobility by itself predisposes to osteoarthritis.

Generalized hypermobility is not usually associated with any obvious disease, but severe laxity is a feature of certain rare connective tissue disorders such as Marfan's syndrome, Ehlers–Danlos syndrome, Larsen's disease and osteogenesis imperfecta.

Deformity

The boundary between variations of the normal and physical deformity is blurred. Indeed, in the development of species, what at one point of time might have been seen as a deformity could over the ages have turned out to be so advantageous as to become essential for survival. So too in humans. The word 'deformity' is derived from the Latin for 'misshapen', but the range of 'normal shape' is so wide that variations should not automatically be designated as deformities, and some undoubted 'deformities' are not necessarily pathological; for example, the generally accepted cut-off points for 'abnormal' shortness or tallness are arbitrary and people who in one population might be considered abnormally short or abnormally tall could, in other populations, be seen as quite ordinary. However, if one leg is short and the other long, no one would quibble with the use of the word 'deformity'!

Specific terms are used to describe the 'position' and 'shape' of the bones and joints. Whether, in any particular case, these amount to 'deformity' will be determined by additional factors such as the extent

to which they deviate from the norm, symptoms to which they give rise, the presence or absence of instability and the degree to which they interfere with function.

Varus and valgus It seems pedantic to replace 'bow legs' and 'knock knees' with 'genu varum' and 'genu valgum', but comparable colloquialisms are not available for deformities of the elbow, hip or big toe; and, besides, the formality is justified by the need for clarity and consistency. Varus means that the part distal to the joint in question is displaced towards the median plane, valgus away from it (Figure 1.11).

Kyphosis and lordosis Seen from the side, the normal spine has a series of curves: convex posteriorly in the thoracic region (kyphosis), and convex anteriorly in the cervical and lumbar regions (lordosis). Excessive curvature constitutes kyphotic or lordotic deformity (also sometimes referred to as hyperkyphosis and hyperlordosis). Colloquially speaking, excessive thoracic kyphosis is referred to as 'round-shouldered'.

Scoliosis Seen from behind, the spine is straight. Any curvature in the coronal plane is called scoliosis. The position and direction of the curve are specified by terms such as thoracic scoliosis, lumbar scoliosis, convex to the right, concave to the left, etc.

Postural deformity A postural deformity is one which the patient can, if properly instructed, correct voluntarily: e.g. thoracic 'kyphosis' due to slumped shoulders. Postural deformity may also be caused by temporary muscle spasm.

Structural deformity A deformity which results from a permanent change in anatomical structure cannot be voluntarily corrected. It is important to distinguish postural scoliosis from structural (fixed) scoliosis. The former is non-progressive and benign; the latter is usually progressive and may require treatment.

'Fixed deformity' This term is ambiguous. It seems to mean that a joint is deformed and unable to move but this is not so. It means that one particular movement cannot be completed. Thus the knee may be able to flex fully but not extend fully – at the limit of its extension it is still 'fixed' in a certain amount of flexion. This would be called a 'fixed flexion deformity'.

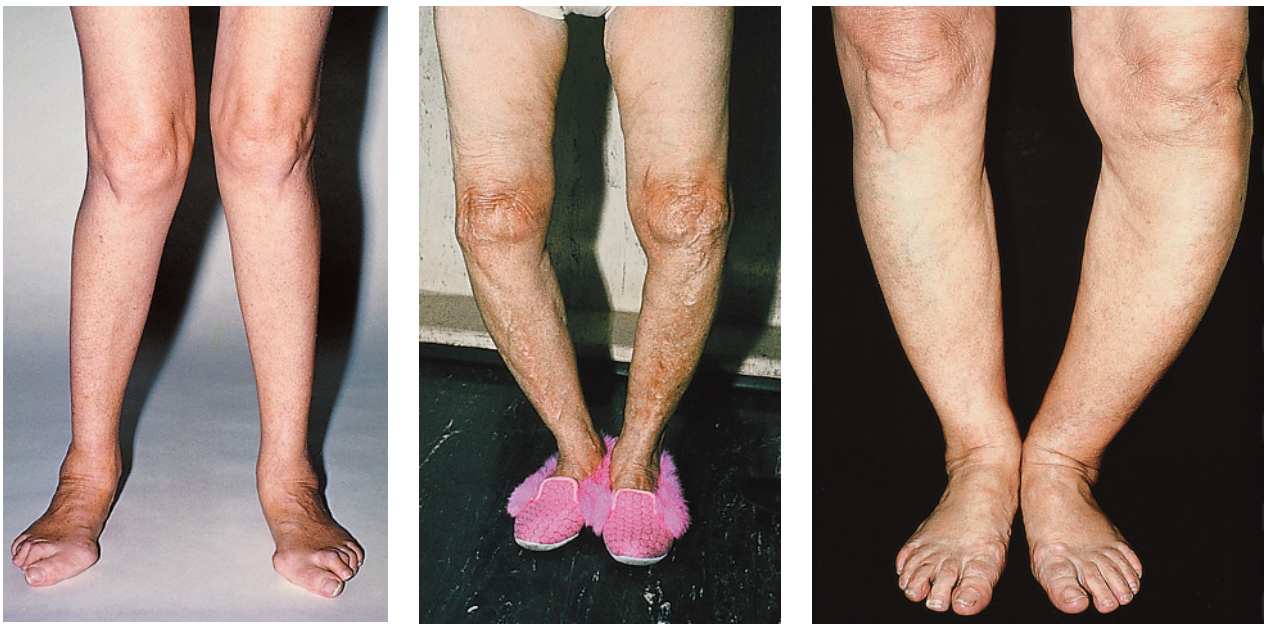
CAUSES OF JOINT DEFORMITY

There are six basic causes of joint deformity.

Contracture of the overlying skin This is seen typically when there is severe scarring across the flexor aspect of a joint, e.g. due to a burn or following surgery.

Contracture of the subcutaneous fascia The classical example is Dupuytren's contracture in the palm of the hand.

Muscle contracture Fibrosis and contracture of muscles that cross a joint will cause a fixed deformity of the joint. This may be due to deep infection or fibrosis following ischaemic necrosis (Volkmann's ischaemic contracture).



(a) (b) (c)

FIgure 1.11 Varus and valgus (a) Valgus knees in a patient with rheumatoid arthritis. The toe joints are also valgus. (b) Varus knees due to osteoarthritis. (c) Another varus knee? No – the deformity here is in the left tibia due to Paget's disease.

Muscle imbalance Unbalanced muscle weakness or spasticity will result in joint deformity which, if not corrected, will eventually become fixed. This is seen most typically in poliomyelitis and cerebral palsy. Tendon rupture, likewise, may cause deformity.

Joint instability Any unstable joint will assume a 'deformed' position when subjected to force.

Joint destruction Trauma, infection or arthritis may destroy the joint and lead to severe deformity.

CAUSES OF BONE DEFORMITY

Bone deformities in small children are usually due to genetic or developmental disorders of cartilage and bone growth; some can be diagnosed *in utero* by special imaging techniques (e.g. achondroplasia); some become apparent when the child starts to walk, or later still during one of the growth spurts (e.g. hereditary multiple exostosis); and some only in early adulthood (e.g. multiple epiphyseal dysplasia). There are a myriad genetic disorders affecting the skeleton, yet any one of these conditions is rare. The least unusual of them are described in Chapter 8.

Acquired deformities in children may be due to fractures involving the physis (growth plate); ask about previous injuries. Other causes include rickets, endocrine disorders, malunited diaphyseal fractures and tumours.

Acquired deformities of bone in adults are usually the result of previous malunited fractures. However, causes such as osteomalacia, bone tumours and Paget's disease should always be considered.

Bony lumps

A bony lump may be due to faulty development, injury, inflammation or a tumour. Although X-ray examination is essential, the clinical features can be highly informative (for example, see Figure 1.12).

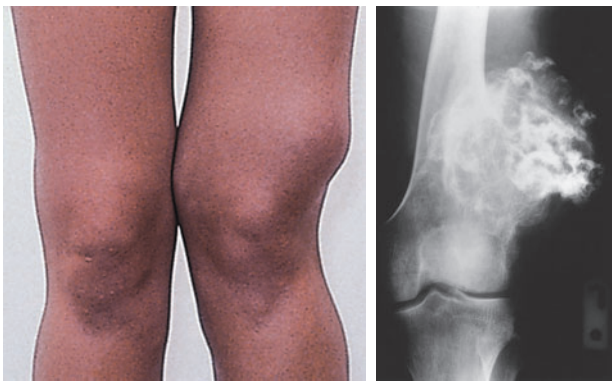


FIgure 1.12 Bony lumps The lump above the left knee is hard, well defined and not increasing in size. The clinical diagnosis of cartilage-capped exostosis (osteochondroma) is confirmed by the X-rays.

Size A large lump attached to bone, or a lump that is getting bigger, is nearly always a tumour.

Site A lump near a joint is most likely to be a tumour (benign or malignant); a lump in the shaft may be fracture callus, inflammatory new bone or a tumour. A benign tumour has a well-defined margin; malignant tumours, inflammatory lumps and callus have a vague edge.

Consistency A benign tumour feels bony and hard; malignant tumours often give the impression that they can be indented.

Tenderness Lumps due to active inflammation, recent callus or a rapidly growing sarcoma are tender.

Multiplicity Multiple bony lumps are uncommon: they occur in hereditary multiple exostosis and in Ollier's disease.

Joint stiffness

The term 'stiffness' covers a variety of limitations. We consider three types of stiffness in particular: (1) all movements absent; (2) all movements limited; (3) one or two movements limited.

All movements absent Surprisingly, although movement is completely blocked, the patient may retain such good function that the restriction goes unnoticed until the joint is examined. Surgical fusion is called 'arthrodesis'; pathological fusion is called 'ankylosis'. Acute suppurative arthritis typically ends in bony ankylosis; tuberculous arthritis heals by fibrosis and causes fibrous ankylosis – not strictly a 'fusion' because there may still be a small jog of movement.

All movements limited After severe injury, movement may be limited as a result of oedema and bruising. Later, adhesions and loss of muscle extensibility may perpetuate the stiffness.

With active inflammation all movements are restricted and painful and the joint is said to be 'irritable'. In acute arthritis spasm may prevent all but a few degrees of movement.

In osteoarthritis the capsule fibroses and movements become increasingly restricted, but pain occurs only at the extremes of motion.

Some movements limited When one particular movement suddenly becomes blocked, the cause is usually mechanical. Thus a torn and displaced meniscus may prevent extension of the knee but not flexion.

Bone deformity may alter the arc of movement, such that it is limited in one direction (loss of abduction in coxa vara is an example) but movement in the opposite direction is full or even increased.

These are all examples of 'fixed deformity'.

DIAGNOSTIC IMAGING

The map is not the territory

Alfred Korzybski

Plain film radiography

Plain film X-ray examination is over 100 years old. Notwithstanding the extraordinary technical advances of the last few decades, it remains the most useful method of diagnostic imaging. Whereas other methods may define an inaccessible anatomical structure more accurately, or may reveal some localized tissue change, the plain film provides information simultaneously on the size, shape, tissue 'density' and bone architecture – characteristics which, taken together, will usually suggest a diagnosis, or at least a range of possible diagnoses.

The radiographic image

X-rays are produced by firing electrons at high speed onto a rotating anode. The resulting beam of X-rays is attenuated by the patient's soft tissues and bones, casting what are effectively 'shadows' which are displayed as images on an appropriately sensitized plate or stored as digital information which is then available to be transferred throughout the local information technology (IT) network. See Figure 1.13.

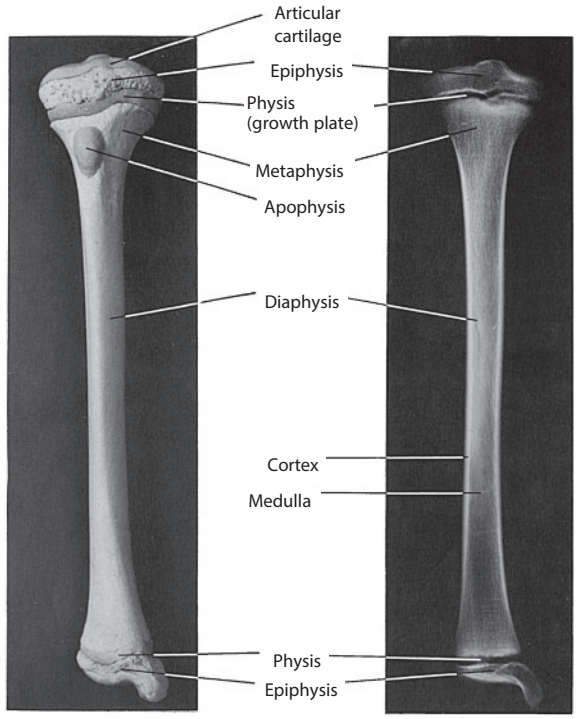


FIgure 1.13 The radiographic image X-ray of an anatomical specimen to show the appearance of various parts of the bone in the X-ray image.

The more dense and impenetrable the tissue, the greater the X-ray attenuation and therefore the more blank, or white, the image that is captured. Thus, a metal implant appears intensely white, bone less so and soft tissues in varying shades of grey depending on their 'density'. Cartilage, which causes little attenuation, appears as a dark area between adjacent bone ends; this 'gap' is usually called the joint space, though of course it is not a space at all, merely a radiolucent zone filled with cartilage. Other 'radiolucent' areas are produced by fluid-filled cysts in bone.

One bone overlying another (e.g. the femoral head inside the acetabular socket) produces superimposed images; any abnormality seen in the resulting combined image could be in either bone, so it is important to obtain several images from different projections in order to separate the anatomical outlines. Similarly, the bright image of a metallic foreign body superimposed upon that of, say, the femoral condyles could mean that the foreign body is in front of, inside or behind the bone. A second projection, at right angles to the first, will give the answer.

Picture Archiving and Communication System (PACS) This is the system whereby all digitally coded images are filed, stored and retrieved to enable the images to be sent to work stations throughout the hospital, to other hospitals or to the Consultant's personal computer.

Radiographic interpretation

Although *radiograph* is the correct word for the plain image which we address, in the present book we have chosen to retain the old-fashioned term 'X-ray', which has become entrenched by long usage. The process of interpreting this image should be as methodical as clinical examination. It is seductively easy to be led astray by some flagrant anomaly; systematic study is the only safeguard. A convenient sequence for examination is: *the patient – the soft tissues – the bones – the joints.*

THE PATIENT

Make sure that the name on the film is that of your patient; mistaken identity is a potent source of error. The clinical details are important; it is surprising how much more you can see on the X-ray when you know the background. Similarly, when requesting an X-ray examination, give the radiologist enough information from the patient's history and the clinical findings to help in guiding his or her thoughts towards the diagnostic possibilities and options. For example, when considering a malignant bone lesion, simply knowing the patient's age may provide an important clue: under the age of 10 it is most likely to be a Ewing's sarcoma; between 10 and 20 years it is more likely to

be an osteosarcoma; and over the age of 50 years it is likely to be a metastatic deposit.

THE SOFT TISSUES

Generalized change Muscle planes are often visible and may reveal wasting or swelling. Bulging outlines around a hip, for example, may suggest a joint effusion; and soft-tissue swelling around interphalangeal joints may be the first radiographic sign of rheumatoid arthritis. Tumours tend to displace fascial planes, whereas infection tends to obliterate them.

Localized change Is there a mass, soft tissue calcification, ossification, gas (from penetrating wound or gas-forming organism) or the presence of a radio-opaque foreign body?

THE BONES

Shape The bones are well enough defined to allow one to check their general anatomy and individual shape (Figure 1.14). For example, for the spine, look at the overall vertebral alignment, then at the disc spaces, and then at each vertebra separately, moving from the body to the pedicles, the facet joints and finally the spinous appendages. For the pelvis, see if the shape is symmetrical with the bones in their normal positions, then look at the sacrum, the two innominate bones, the pubic rami and the ischial tuberosities, then the femoral heads and the upper ends of the femora, always comparing the two sides.

Generalized change Take note of changes in bone 'density' (osteopaenia or osteosclerosis). Is there abnormal trabeculation, as in Paget's disease (Figure 1.15a)? Are there features suggestive of diffuse metastatic infiltration, either sclerotic or lytic? Other polyostotic lesions include fibrous dysplasia, histiocyotis, multiple exostosis and Paget's disease. With aggressive-looking polyostotic lesions, think of metastases (including myeloma and lymphoma) and also multifocal infection. By contrast, most primary tumours are monostotic.

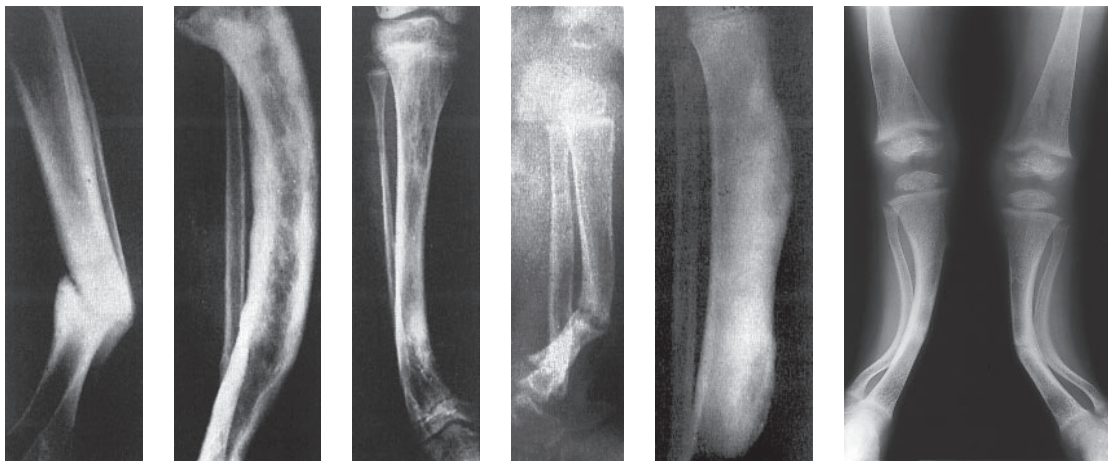
Localized change Focal abnormalities should be approached in the same way as one would conduct a clinical analysis of a soft tissue abnormality. Start describing the abnormality from the centre and move outwards. Determine the lesion's size, site, shape, density and margins, as well as adjacent periosteal changes and any surrounding soft tissue changes. Remember that benign lesions are usually well defined with sclerotic margins (Figure 1.15b) and a smooth periosteal reaction. Ill-defined areas with permeative bone destruction (Figure 1.15c) and irregular or speculated periosteal reactions (Figure 1.15d) suggest an aggressive lesion such as infection or a malignant tumour.

THE JOINTS

The radiographic 'joint' consists of the articulating bones and the 'space' between them.

The 'joint space' The joint space is, of course, illusory; it is occupied by a film of synovial fluid plus radiolucent articular cartilage which varies in thickness from 1 mm or less (the carpal joints) to 6 mm (the knee). It looks much wider in children than in adults because much of the epiphysis is still cartilaginous and therefore radiolucent. Lines of increased density within the radiographic articular 'space' may be due to calcification of the cartilage or menisci (chondrocalcinosis). Loose bodies, if they are radio-opaque, appear as rounded patches overlying the normal structures.

Shape Note the general orientation of the joint and the congruity of the bone ends (actually the subarticular bone plates), if necessary comparing the



(a) (b) (c) (d) (e) (f)

FIgure 1.14 X-rays – bent bones (a) Malunited fracture; (b) Paget's disease; (c) dyschondroplasia; (d) congenital pseudarthrosis; (e) syphilitic sabre tibia; (f) osteogenesis imperfecta.

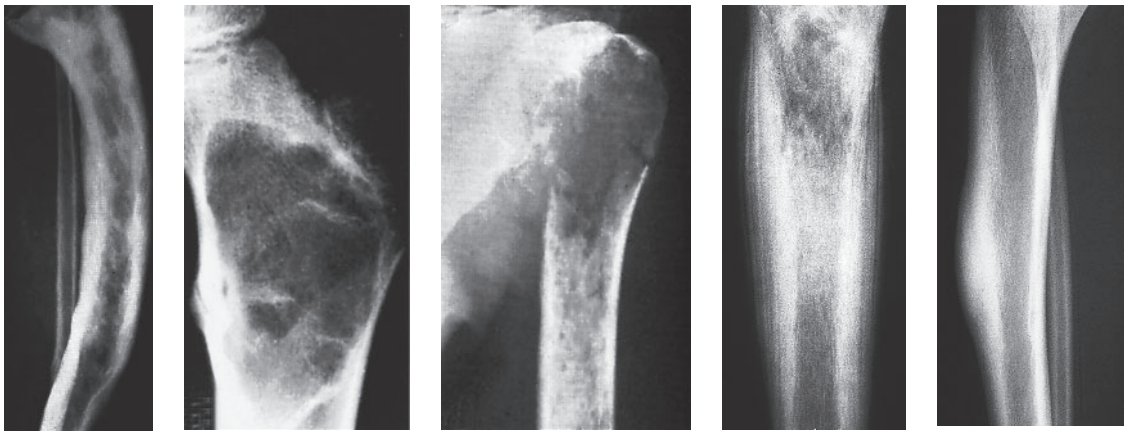


FIgure 1.15 X-rays – important features to look for (a) *General shape and appearance*, in this case the cortices are thickened and the bone is bent (Paget's disease). (b,c) *Interior density*, a vacant area may represent a true cyst (b), or radiolucent material infiltrating the bone, like the metastatic tumour in (c). (d) *Periosteal reaction*, typically seen in healing fractures, bone infection and malignant bone tumours – as in this example of Ewing's sarcoma. Compare this with the smooth periosteal new bone formation shown in (e).

abnormal with the normal opposite side. Then look for narrowing or asymmetry of the joint 'space': narrowing signifies loss of hyaline cartilage and is typical of infection, inflammatory arthropathies and osteoarthritis. Further stages of joint destruction are revealed by irregularity of the radiographically visible bone ends and radiolucent cysts in the subchondral bone. Bony excrescences at the joint margins (osteophytes) are typical of osteoarthritis.

Erosions Look for associated bone erosions. The position of erosions and symmetry help to define various types of arthropathy. In rheumatoid arthritis and psoriasis the erosions are periarticular (at the bare area where the hyaline cartilage covering the joint has ended and the intracapsular bone is exposed to joint fluid). In gout the erosions are further away from the articular surfaces and are described as juxta-articular. Rheumatoid arthritis is classically symmetrical and predominantly involves the metacarpophalangeal and proximal interphalangeal joints in both hands. The erosions in psoriasis are usually more feathery with ill-defined new bone at their margins. Ill-defined erosions suggest active synovitis whereas corticated erosions indicate healing and chronicity.

Diagnostic associations

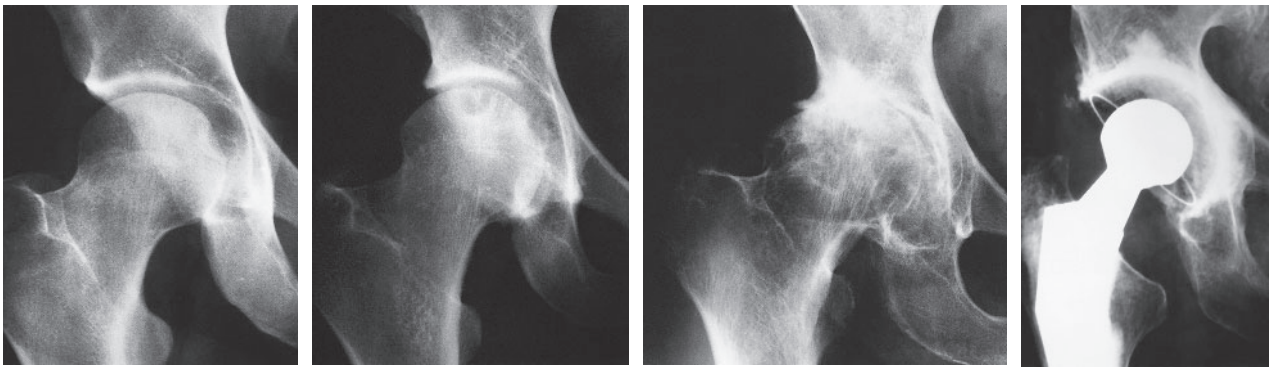
However carefully the individual X-ray features are observed, the diagnosis will not leap ready-made off the X-ray plate. Even a fracture is not always obvious. It is the pattern of abnormalities that counts: if you see one feature that is suggestive, look for others that are commonly associated.

- Narrowing of the joint space + subchondral sclerosis and cysts + osteophytes = osteoarthritis (Figure 1.16).
- Narrowing of the joint space + osteoporosis + periarticular erosions = inflammatory arthritis. Add to this the typical distribution, more or less symmetrically in the proximal joints of both hands, and you must think of rheumatoid arthritis.
- Bone destruction + periosteal new bone formation = infection or malignancy until proven otherwise.
- Remember: the next best investigation is either the previous radiograph or the subsequent follow-up radiograph. Sequential films demonstrate either progression of changes in active pathology or status quo in long-standing conditions.

Limitations of conventional radiography

Conventional radiography involves exposure of the patient to ionizing radiation, which under certain circumstances can lead to radiation-induced cancer. The Ionizing Radiation Medical Exposure Regulations (IRMER) 2000 are embedded in European Law, requiring all clinicians to justify any exposure of the patient to ionizing radiation. It is a criminal offence to breach these regulations. Ionizing radiation can also damage a developing fetus, especially in the first trimester.

As a diagnostic tool, conventional radiography provides poor soft-tissue contrast: for example, it cannot distinguish between muscles, tendons, ligaments and hyaline cartilage. Ultrasound (US), computed tomography (CT) and magnetic resonance imaging (MRI) are now employed to complement plain X-ray examination. However, in parts of the world where these techniques are not available, some modifications of plain radiography still have a useful role.



(a) (b) (c) (d)

Figure 1.16 Plain X-rays of the hip Stages in the development of osteoarthritis (OA). (a) Normal hip: anatomical shape and position, with joint 'space' (articular cartilage) fully preserved. (b) Early OA, showing joint space slightly decreased and a subarticular cyst in the femoral head. (c) Advanced OA: joint space markedly decreased; osteophytes at the joint margin. (d) Hip replacement: the cup is radiolucent but its position is shown by a circumferential wire marker. Note the differing image 'densities': (1) the metal femoral implant; (2) the polyethylene cup (radiolucent); (3) acrylic cement impacted into the adjacent bone.

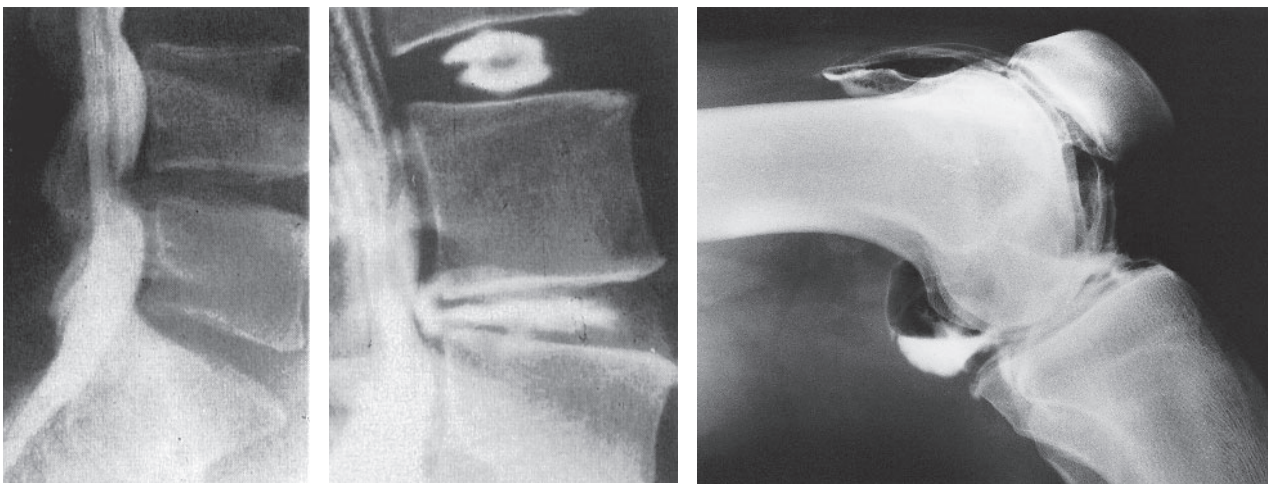
X-RAYS USING CONTRAST MEDIA

Substances that alter X-ray attenuation characteristics can be used to produce images which contrast with those of the normal tissues. The contrast media used in orthopaedics are mostly iodine-based liquids which can be injected into sinuses, joint cavities or the spinal theca (Figure 1.17). Air or gas also can be injected into joints to produce a 'negative image' outlining the joint cavity.

Oily iodides are not absorbed and maintain maximum concentration after injection. However, because they are non-miscible, they do not penetrate well into all the nooks and crannies. They are also tissue irritants, especially if used intrathecally and are now rarely used as they have been shown to cause adhesive arachnoiditis. Ionic, water-soluble iodides permit much more detailed imaging and, although also somewhat irritant and neurotoxic, are rapidly absorbed and excreted.

Sinography

Sinography is the simplest form of contrast radiography. The medium (usually one of the ionic water-soluble compounds) is injected into an open sinus; the film shows the track and whether or not it leads to the underlying bone or joint.



(a) (b) (c)

Figure 1.17 Contrast radiography (a) Myelography shows the outline of the spinal theca. Where facilities are available, myelography has been largely replaced by CT and MRI. (b) Discography is sometimes useful: note the difference between a normal intervertebral disc (upper level) and a degenerate disc (lower level). (c) Contrast arthrography of the knee shows a small popliteal herniation.

Arthrography

Arthrography is a particularly useful form of contrast radiography. Intra-articular loose bodies will produce filling defects in the opaque contrast medium. In the knee, torn menisci, ligament tears and capsular ruptures can be shown. In children's hips, arthrography is a useful method of outlining the cartilaginous (and therefore radiolucent) femoral head. In adults with avascular necrosis of the femoral head, arthrography may show up torn flaps of cartilage. After hip replacement, loosening of a prosthesis may be revealed by seepage of the contrast medium into the cement/bone interface. In the hip, ankle, wrist and shoulder, the injected contrast medium may disclose labral tears or defects in the capsular structures. In the spine, contrast radiography can be used to diagnose disc degeneration (discography) and abnormalities of the small facet joints (facetography).

Myelography

Myelography was used extensively in the past for the diagnosis of disc prolapse and other spinal canal lesions. It has been largely replaced by non-invasive methods such as CT and MRI. However, it still has a place in the investigation of nerve root lesions and as an adjunct to other methods in patients with back pain.

The oily media are no longer used, and even with the ionic water-soluble iodides there is a considerable incidence of complications, such as low-pressure headache (due to the lumbar puncture), muscular spasms or convulsions (due to neurotoxicity, especially if the chemical is allowed to flow above the mid-dorsal region) and arachnoiditis (which is attributed to the hyperosmolality of these compounds in relation to cerebrospinal fluid). Precautions, such as keeping the patient sitting upright after myelography, must be strictly observed.

Metrizamide has low neurotoxicity and at working concentrations it is more or less isotonic with cerebrospinal fluid. It can therefore be used throughout the length of the spinal canal; the nerve roots are also well delineated (radiculography). A bulging disc, an intrathecal tumour or narrowing of the bony canal will produce characteristic distortions of the opaque column in the myelogram.

Plain tomography

Tomography provides an image 'focused' on a selected plane. By moving the tube and the X-ray film in opposite directions around the patient during the exposure, images on either side of the pivotal plane are deliberately blurred out. When several 'cuts' are studied, lesions obscured in conventional X-rays may be revealed. The method is useful for diagnosing segmental bone necrosis and depressed fractures in cancellous bone (e.g. of the vertebral body or the tibial plateau); these defects are often obscured in the plain X-ray by the surrounding intact mass of bone. Small radiolucent lesions, such as osteoid osteomas and bone abscesses, can also be revealed.

A useful procedure in former years, conventional tomography has been largely supplanted by CT and MRI.

Computed tomography (CT)

Like plain tomography, CT produces sectional images through selected tissue planes – but with much greater resolution (Figure 1.18). A further advance over conventional tomography is that the images are trans-axial (like transverse anatomical sections), thus exposing anatomical planes that are never viewed in plain film X-rays. A general (or 'localization') view is obtained, the region of interest is selected and a series of cross-sectional images is produced and digitally recorded. 'Slices' through the larger joints or tissue masses may be 3–5 mm apart; those through the small joints or intervertebral discs have to be much thinner.

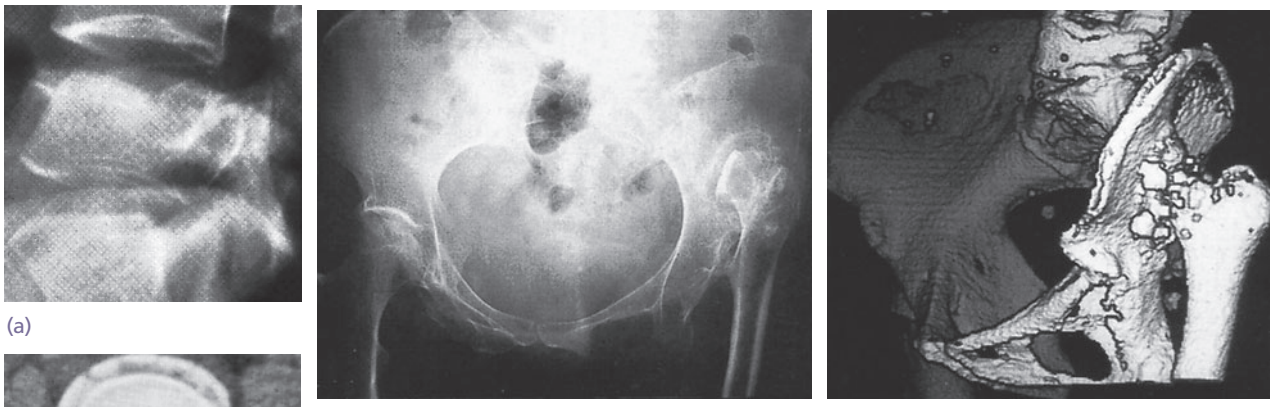
New multislice CT scanners provide images of high quality from which multiplanar reconstructions in all three orthogonal planes can be produced. Three-dimensional surface rendered reconstructions and volume rendered reconstructions may help in demonstrating anatomical contours, but fine detail is lost in this process.

Clinical applications

Because CT achieves excellent contrast resolution and spatial localization, it is able to display the size, shape and position of bone and soft-tissue masses in transverse planes. Image acquisition is extremely fast. The technique is therefore ideal for evaluating acute trauma to the head, spine, chest, abdomen and pelvis. It is better than MRI for demonstrating fine bone detail and soft-tissue calcification or ossification.

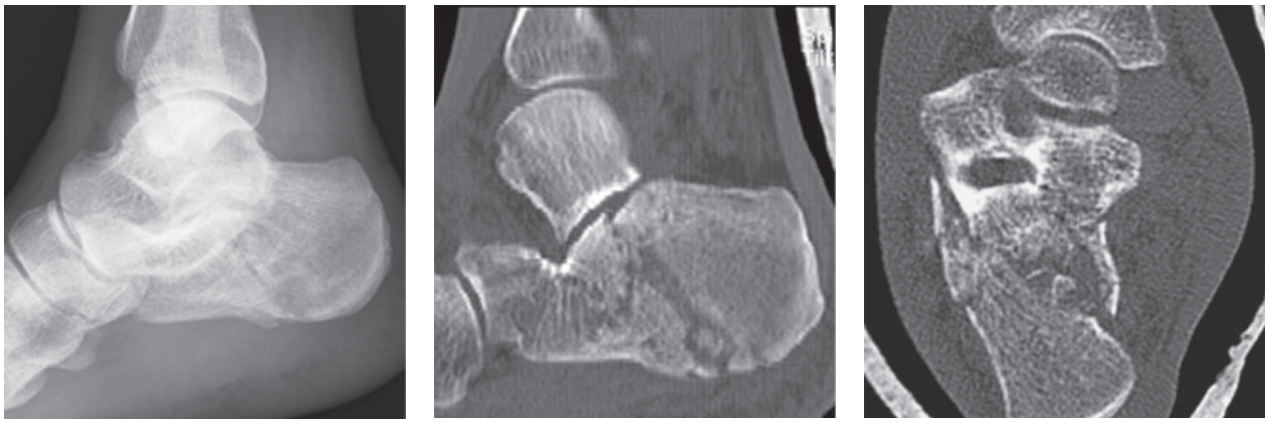
Computed tomography is also an invaluable tool for assisting with preoperative planning in secondary fracture management. It is routinely used for assessing injuries of the vertebrae, acetabulum, proximal tibial plateau, ankle and foot – indeed complex fractures and fracture-dislocations at any site (Figure 1.19).

It is also useful in the assessment of bone tumour size and spread, even if it is unable to characterize the tumour type. It can be employed for guiding soft-tissue and bone biopsies.



(a) (b) (c) (d)

Figure 1.18 Computed tomography (CT) The plain X-ray (a) shows a fracture of the vertebral body but one cannot tell precisely how the bone fragments are displaced. The CT (b) shows clearly that they are dangerously close to the cauda equina. (c) Congenital hip dislocation, defined more clearly by (d) three-dimensional CT reconstruction.



(a) (b) (c)

Figure 1.19 CT for complex fractures (a) A plain X-ray shows a fracture of the calcaneum but the details are obscure. CT sagittal and axial views (b,c) give a much clearer idea of the seriousness of this fracture.

Limitations

An important limitation of CT is that it provides relatively poor soft-tissue contrast when compared with MRI.

A major disadvantage of this technique is the relatively high radiation exposure to which the patient is subjected. It should, therefore, be used with discretion.

Magnetic resonance imaging (MRI)

Magnetic resonance imaging produces cross-sectional images of any body part in any plane. It yields superb soft-tissue contrast, allowing different soft tissues to be clearly distinguished, e.g. ligaments, tendons, muscle and hyaline cartilage. Another big advantage of MRI is that it does not use ionizing radiation. It is, however, contraindicated in patients with pacemakers and possible metallic foreign bodies in the eye or brain, as these could potentially move when the patient is introduced into the scanner's strong magnetic field. Approximately 5% of patients cannot tolerate the scan due to claustrophobia, but newer scanners are being developed to be more 'open'.

MRI physics

The patient's body is placed in a strong magnetic field (between 5 and 30 000 times the strength of the Earth's magnetic field). The body's protons have a positive charge and align themselves along this strong

external magnetic field. The protons are spinning and can be further excited by radiofrequency pulses, rather like whipping a spinning top. These spinning positive charges will not only induce a small magnetic field of their own, but will produce a signal as they relax (slow down) at different rates.

A proton density map is recorded from these signals and plotted in *x*, *y* and *z* coordinates. Different speeds of tissue excitation with radiofrequency pulses (repetition times, or TR) and different intervals between recording these signals (time to echo, or TE) will yield anatomical pictures with varying 'weighting' and characteristics. T1 weighted (T1W) images have a high spatial resolution and provide good anatomical-looking pictures. T2 weighted (T2W) images give more information about the physiological characteristics of the tissue. Proton density (PD) images are also described as 'balanced' or 'intermediate' as they are essentially a combination of T1 and T2 weighting and yield excellent anatomical detail for orthopaedic imaging. Fat suppression sequences allow highlighting of abnormal water, which is particularly useful in orthopaedics when assessing both soft-tissue and bone marrow oedema.

Intravenous contrast

Just as in CT, enhancement by intravenous contrast relies on an active blood supply and leaky cell membranes. Areas of inflammation and active tumour tissue will be highlighted. Gadolinium compounds are employed as they have seven unpaired electrons and work by creating local magnetic field disturbances at their sites of accumulation.

Indirect arthrography

Gadolinium compounds administered intravenously will be secreted through joint synovium into joint effusions resulting in indirect arthrography. However, there is no additional distension of the joint, which limits its effect.

Direct arthrography

Direct puncture of joints under image guidance with a solution containing dilute gadolinium (1:200 concentration) is routinely performed. This provides a positive contrast within the joint and distension of the joint capsule, thereby separating many of the closely applied soft-tissue structures that can be demonstrated on the subsequent MRI scan.

Clinical applications

Magnetic resonance imaging is becoming cheaper and more widely available. Its excellent anatomical detail, soft-tissue contrast and multiplanar capability make it ideal for non-invasive imaging of the musculoskeletal system (Figures 1.20 and 1.21). The multiplanar capability provides accurate cross-sectional information and the axial images in particular will reveal detailed limb compartmental anatomy. The excellent soft-tissue contrast allows identification of similar density soft tissues, for example in distinguishing between tendons, cartilage and ligaments. By using combinations of T1W, T2W and fat-suppressed sequences, specific abnormalities can be further characterized with tissue specificity, so further extending the diagnostic possibilities.

In orthopaedic surgery, MRI of the hip, knee, ankle, shoulder and wrist is now fairly commonplace. It can detect the early changes of bone marrow oedema and osteonecrosis before any other imaging

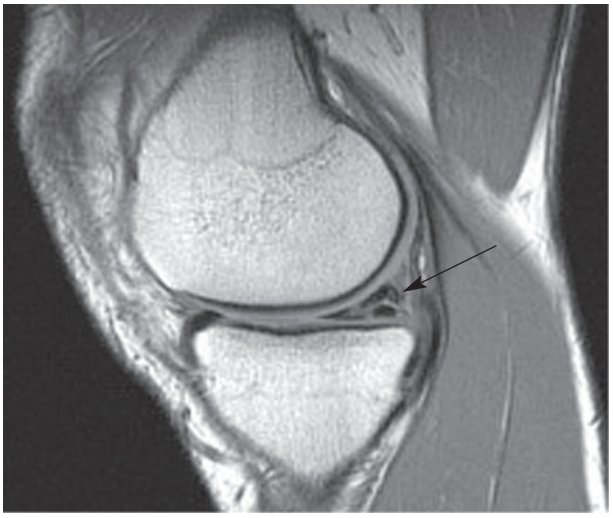
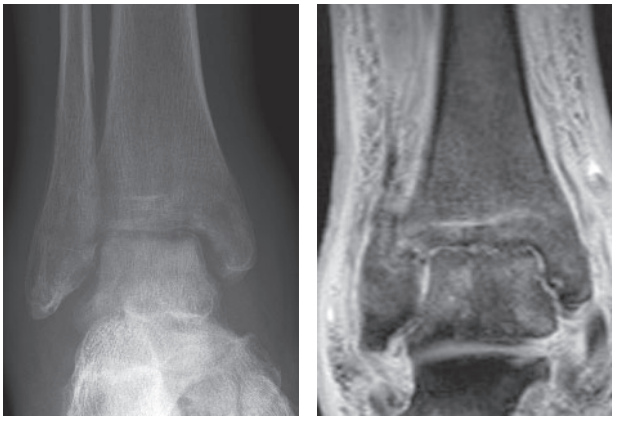


Figure 1.20 Magnetic resonance imaging MRI is ideal for displaying soft-tissue injuries, particularly tears of the menisci of the knee; this common injury is clearly shown in the picture.



(a) (b)

Figure 1.21 MRI A case of septic arthritis of the ankle, suspected from the plain X-ray (a) and confirmed by MRI (b).

modality. In the knee, MRI is as accurate as arthroscopy in diagnosing meniscal tears and cruciate ligament injuries. Bone and soft-tissue tumours should be routinely examined by MRI as the intraosseous and extraosseous extent and spread of disease, as well as the compartmental anatomy, can be accurately assessed. Additional use of fat-suppression sequences determines the extent of perilesional oedema and intravenous contrast will demonstrate the active part of the tumour.

Intravenous contrast is used to distinguish vascularized from avascular tissue (e.g. following a scaphoid fracture) or in defining active necrotic areas of tumour, or in demonstrating areas of active inflammation.

Direct MRI arthrography is used to distend the joint capsule and outline labral tears in the shoulder and the hip. In the ankle, it provides the way to demonstrate anterolateral impingement and assess the integrity of the capsular ligaments.

New generation MRI scanners

Many new scanners are being developed in the clinical setting using more powerful magnetic fields. Previously, the field strength was commonly between 0.5 and 1.5 Tesla. More recently, scanners using 3 Tesla have started being introduced. The increased field strength yields improved contrast and definition, but it is also more susceptible to artefacts.

Dedicated small-part scanners are also being introduced to assess limbs, for example for occult scaphoid fractures in the Emergency Department. Upright scanners have been developed to assess pathology that is apparent only when the patient is weight-bearing.

Limitations

Despite its undoubted value, MRI (like all singular methods of investigation) has its limitations and it must be seen as one of a group of imaging modalities, none of which by itself is appropriate in every situation. Conventional radiographs and CT are more sensitive to soft-tissue calcification and ossification, changes which can easily be easily overlooked on MRI. Conventional radiographs should therefore be used in combination with MRI to prevent such errors.

Diagnostic ultrasound

High-frequency sound waves, generated by a transducer, can penetrate several centimetres into the soft tissues; as they pass through the tissue interfaces, some of these waves are reflected back (like echoes) to the transducer, where they are registered as electrical signals and displayed as images on a screen. Unlike X-rays, the image does not depend on tissue density but rather on reflective surfaces and soft-tissue interfaces. This is the same principle as applies in sonar detection for ships or submarines.

Depending on their structure, different tissues are referred to as highly echogenic, mildly echogenic or echo-free. Fluid-filled cysts are echo-free; fat is highly echogenic; and semi-solid organs manifest varying degrees of 'echogenicity', which makes it possible to differentiate between them.

Real-time display on a monitor gives a dynamic image, which is more useful than the usual static images. A big advantage of this technique is that the equipment is simple and portable and can be used almost anywhere; another is that it is entirely harmless.

Clinical applications

Because of the marked echogenic contrast between cystic and solid masses, ultrasonography is particularly useful for identifying hidden 'cystic' lesions such as haematomas, abscesses, popliteal cysts and arterial aneurysms. It is also capable of detecting intra-articular fluid and may be used to diagnose a synovial effusion or to monitor the progress of an 'irritable hip'.

Ultrasound is commonly used for assessing tendons and diagnosing conditions such as tendinitis and partial or complete tears. The rotator cuff, patellar ligament, quadriceps tendon, Achilles tendon, flexor tendons and peroneal tendons are typical examples. The same technique is used extensively for guiding needle placement in diagnostic and therapeutic joint and soft-tissue injections. Another important application is in the screening of newborn babies for congenital dislocation (or dysplasia) of the hip; the cartilaginous femoral head and acetabulum (which are, of course, 'invisible' on X-ray) can be clearly identified, and their relationship to each other shows whether the hip is normal or abnormal.

Ultrasound imaging is quick, cheap, simple and readily available. However, the information obtained is highly operator-dependent, relying on the experience and interpretation of the technician.

Doppler ultrasound

Blood flow can be detected by using the principle of a change in frequency of sound when material is moving towards or away from the ultrasound transducer. This is the same principle as the change in frequency of the noise from a passing fire engine when travelling towards and then away from an observer. Abnormal increased blood flow can be observed in areas of inflammation or in aggressive tumours. Different flow rates can be shown by different colour representations ('colour Doppler').

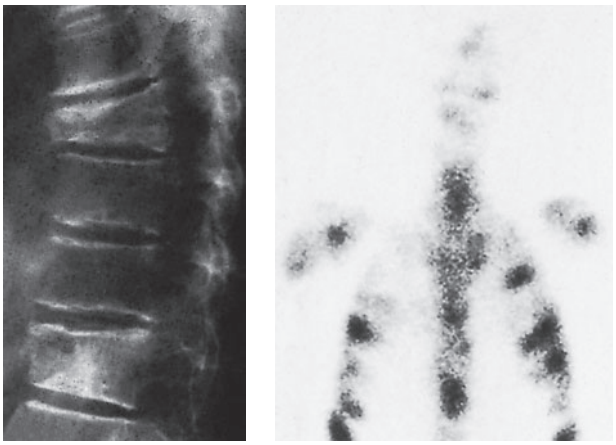
Radionuclide imaging

Photon emission by radionuclides taken up in specific tissues can be recorded by a gamma camera to produce an image which reflects physiological activity in that tissue or organ. The radiopharmaceutical used for radionuclide imaging has two components: a chemical compound that is chosen for its metabolic uptake in the target tissue or organ, and a radioisotope tracer that will emit photons for detection.

Isotope bone scans

For bone imaging, the ideal isotope is technetium-99m (^{99m}Tc): it has the appropriate energy characteristics for gamma camera imaging, it has a relatively short half-life (6 hours) and it is rapidly excreted in the urine. A bone-seeking phosphate compound is used as the substrate as it is selectively taken up and concentrated in bone. The low background radioactivity means that any site of increased uptake is readily visible (Figure 1.22).

Technetium-labelled hydroxymethylene diphosphonate (^{99m}Tc-HDP) is injected intravenously and its activity is recorded at two stages: (1) the early perfusion phase, shortly after injection, while the isotope is still in the blood stream or the perivascular space thus reflecting local blood flow difference; and (2) the delayed bone phase, 3 hours later, when the isotope has been taken up in bone tissue. Normally, in the early perfusion phase the vascular soft tissues around the joints produce the sharpest (most active) image; 3 hours later this activity has faded and the bone outlines are shown more clearly, the greatest activity appearing in the cancellous tissue at the ends of the long bones.



(a) (b)

Figure 1.22 Radionuclide scanning (a) The plain X-ray showed a pathological fracture, probably through a metastatic tumour. (b) The bone scan revealed generalized secondaries, here involving the spine and ribs.

Changes in radioactivity are most significant when they are localized or asymmetrical. The following four types of abnormality are seen.

Increased activity in the perfusion phase This is due to increased soft-tissue blood flow, suggesting inflammation (e.g. acute or chronic synovitis), a fracture, a highly vascular tumour or regional sympathetic dystrophy.

Decreased activity in the perfusion phase This is much less common and signifies local vascular insufficiency.

Increased activity in the delayed bone phase This could be due either to excessive isotope uptake in the osseous extracellular fluid or to more avid incorporation into newly forming bone tissue; either would be likely in a fracture, implant loosening, infection, a local tumour or healing after necrosis, and nothing in the bone scan itself distinguishes between these conditions.

Diminished activity in the bone phase This is due to an absent blood supply (e.g. in the femoral head after a fracture of the femoral neck) or to replacement of bone by pathological tissue.

CLINICAL APPLICATIONS

Radionuclide imaging is useful in many situations: (1) the diagnosis of stress fractures or other undisplaced fractures that are not detectable on the plain X-ray; (2) the detection of a small bone abscess, or an osteoid osteoma; (3) the investigation of loosening or infection around prostheses; (4) the diagnosis of femoral head ischaemia in Perthes' disease or avascular necrosis in adults; (5) the early detection of bone metastases. The scintigraphic appearances in these conditions are described in the relevant chapters. In most cases the isotope scan serves chiefly to pinpoint the site of abnormality and it should always be viewed in conjunction with other modes of imaging.

Bone scintigraphy is relatively sensitive but nonspecific. One advantage is that the whole body can be imaged to look for multiple sites of pathology (occult metastases, multifocal infection and multiple occult fractures). It is also one of the only techniques to give information about physiological activity in the tissues being examined (essentially osteoblastic activity). However, the technique carries a significant radiation burden (equivalent to approximately 200 chest X-rays) and the images yielded make anatomical localization difficult (poor spatial resolution). For localized problems MRI has superseded bone scintigraphy as it yields much greater specificity due to its superior anatomical depiction and tissue specificity.

Other radionuclide compounds

Gallium-67 (^{67}Ga) Gallium-67 concentrates in inflammatory cells and has been used to identify sites of hidden infection: for example, in the investigation of prosthetic loosening after joint replacement. However, it is arguable whether it gives any more reliable information than the ^{99m}Tc bone scan.

Indium-111-labelled leucocytes (^{111}I) The patient's own white blood cells are removed and labelled with indium-111 before being re-injected into the patient's bloodstream. Preferential uptake in areas of infection is expected, thereby hoping to distinguish sites of active infection from chronic inflammation. For example, white cell uptake is more likely to be seen with an infected total hip replacement as opposed to mechanical loosening. However, as this technique is expensive and still not completely specific, it is seldom performed.

Single-photon emission computed tomography (SPECT)

Single-photon emission computed tomography (SPECT) is essentially a bone scan in which images are recorded and displayed in all three orthogonal planes. Coronal, sagittal and axial images at multiple levels make spatial localization of pathology possible: for example, activity in one side of a lumbar vertebra on the planar images can be further localized to the body, pedicle or lamina of the vertebra on the SPECT images.

Positron emission tomography (PET)

Positron emission tomography (PET) is an advanced nuclear medicine technique that allows functional imaging of disease processes. Positron-emitting isotopes with short half-lives are produced on site at specialist centres using a cyclotron. Various radiopharmaceuticals can be employed, but currently the most commonly used is 18-fluoro-2-deoxy-D-glucose (^{18}FDG). The ^{18}FDG is accumulated in different parts of the body where it can effectively measure the rate of consumption of glucose. Malignant tumours metabolize glucose at a faster rate than benign tumours and PET scanners are extremely useful in looking for occult sites of disease around the body on this basis.

PET/CT is a hybrid examination performing both PET and CT on the patient in order to superimpose the two images produced. The combination of these two techniques uses the sensitivity of PET for functional tissue changes and the cross-sectional anatomy detail of CT to localize the position of this activity.

PET is useful in oncology to identify occult malignant tumours and metastases and more accurately 'stage' the disease. Furthermore, activity levels at known sites of disease can be used to assess treatment and distinguish 'active' residual tumour or tumour recurrence from 'inactive' post-surgical scarring and necrotic tumour.

Bone mineral densitometry

Bone mineral density (BMD) measurement is now widely used in identifying patients with osteoporosis and an increased risk of osteoporotic fractures. Various techniques have been developed, including radiographic absorptiometry (RA), quantitative computed tomography (QCT) and quantitative ultrasonometry (QUS). However, the most widely used technique is dual energy X-ray absorptiometry (DXA) (Figure 1.23).

RA uses conventional radiographic equipment and measures bone density in the phalanges. QCT measures trabecular bone density in vertebral bodies, but is not widely available and involves a higher dose of ionizing radiation than DXA. QUS assesses bone mineral density in the peripheral skeleton (e.g. the wrist and calcaneus) by measuring both the attenuation of ultrasound and the variation of speed of sound through the bone.

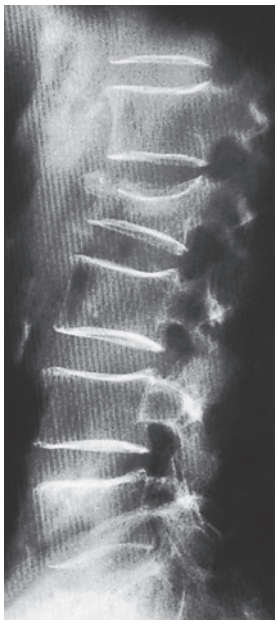
DXA employs columnated low-dose X-ray beams of two different energy levels in order to distinguish the density of bone from that of soft tissue. Although this involves the use of ionizing radiation, it is an extremely low dose. A further advantage of DXA is the development of a huge international database that allows expression of bone mineral density values in comparison both to an age- and sex-matched population (Z score) and also to the peak adult bone mass (T score). The T score in particular allows calculation of relative fracture risk. Individual values for both the lumbar spine and hips are obtained as there is often a discrepancy between these two sites and the fracture risk is more directly related to the value at the target area. By World Health Organization (WHO) criteria, T scores of < -1.0 indicate 'osteopenia' and T scores of < -2.5 indicate 'osteoporosis'.

BLOOD TESTS

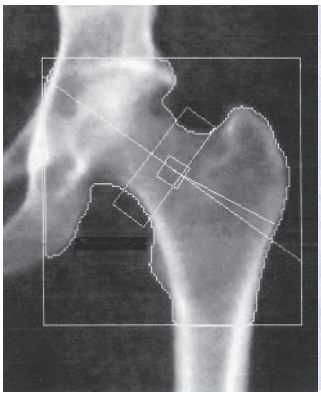
Non-specific blood tests

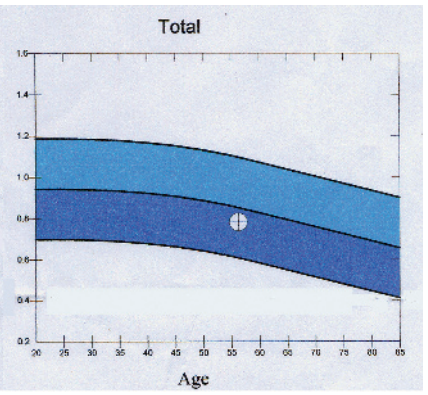
Non-specific blood abnormalities are common in bone and joint disorders; their interpretation hinges on the clinical and X-ray findings.

Hypochromic anaemia This is usual in rheumatoid arthritis, but it may also be a consequence of



(a)





DXA Results Summary:

Region	Area (cm^2)	BMC (g)	BMD (g/cm^2)	T score	PR(%)	Z score	AM(%)
Neck	5.37	3.62	0.675	–1.6	79	–0.5	93
Troch	12.48	7.10	0.569	–1.3	81	–0.6	90
Inter	20.07	18.92	0.943	–1.0	86	–0.5	92
Total	**37.92**	**29.64**	**0.782**	**–1.3**	**83**	**–0.6**	**92**
Ward's	1.03	0.54	0.527	–1.8	72	0.0	100

Total BMD CV 1.)%
WHO Classification: Osteopenia
Fracture Risk: Increased

(b)

Figure 1.23 Measurement of bone mass (a) X-ray of the lumbar spine shows a compression fracture of L2. The general loss of bone density accentuates the cortical outlines of the vertebral body end-plates. These features are characteristic of diminished bone mass, which can be measured accurately by dual energy X-ray absorptiometry. (b) DXA scan from another woman who attended for monitoring at the onset of the menopause.

gastrointestinal bleeding due to the anti-inflammatory drugs.

Leucocytosis Although generally associated with infection, a mild leucocytosis is not uncommon in rheumatoid arthritis and during an attack of gout.

The erythrocyte sedimentation rate (ESR) ESR is usually increased in acute and chronic inflammatory disorders and after tissue injury. However, patients with low-grade infection may have a normal ESR and this should not be taken as a reassuring sign. The ESR is strongly affected by the presence of monoclonal immunoglobulins; a high ESR is almost mandatory in the diagnosis of myelomatosis.

C-reactive protein (and other acute phase proteins) These may be abnormally increased in chronic inflammatory arthritis and (temporarily) after injury or operation. The test is often used to monitor the progress and activity of rheumatoid arthritis and chronic infection.

Plasma gamma globulins Measured by protein electrophoresis. Their precise characterization is helpful in the assessment of certain rheumatic disorders, and more particularly in the diagnosis of myelomatosis.

Rheumatoid factor tests

Rheumatoid factor, an IgM autoantibody, is present in about 75% of adults with rheumatoid arthritis. However, it is not pathognomonic: some patients with undoubted rheumatoid arthritis remain 'seronegative', while rheumatoid factor is found in some patients with other disorders such as systemic lupus erythematosus and scleroderma.

Ankylosing spondylitis, Reiter's disease and psoriatic arthritis characteristically test negative for rheumatoid factor; they have been grouped together as the 'seronegative spondarthritides'.

Tissue typing

Human leucocyte antigens (HLA) can be detected in white blood cells and they are used to characterize individual tissue types. The seronegative spondarthritides are closely associated with the presence of HLAB27 on chromosome 6; this is frequently used as a confirmatory test in patients suspected of having ankylosing spondylitis or Reiter's disease, but it should not be regarded as a specific test because it is positive in about 8% of normal Western Europeans.

Biochemistry

Biochemical tests are essential in monitoring patients after any serious injury. They are also used routinely in the investigation of rheumatic disorders and abnormalities of bone metabolism. Their significance is discussed under the relevant conditions.

SYNOVIAL FLUID ANALYSIS

Arthrocentesis and synovial fluid analysis is a much neglected diagnostic procedure; given the correct indications, it can yield valuable information. It should be considered in the following conditions:

Acute joint swelling after injury The distinction between synovitis and bleeding may not be obvious; aspiration will settle the question immediately.

Acute atraumatic synovitis in adults Synovial fluid analysis may be the only way to distinguish between infection, gout and pseudogout. Characteristic crystals can be identified on polarized light microscopy.

Suspected infection Careful examination and laboratory investigations may provide the answer, but they take time. Joint aspiration is essential for early diagnosis.

Chronic synovitis Here joint aspiration is less urgent, and is only one of many diagnostic procedures in the investigation of suspected tuberculosis or atypical rheumatic disorders.

Technique

Joint aspiration should always be performed under strict aseptic conditions. After infiltrating the skin with a local anaesthetic, a 20-gauge needle is introduced and a sample of joint fluid is aspirated; even a small quantity of fluid (less than 0.5 mL) is enough for diagnostic analysis.

The volume of fluid and its appearance are immediately noted. Normal synovial fluid is clear and slightly yellow. A cloudy or turbid fluid is due to the presence of cells, usually a sign of inflammation. Blood-stained fluid may be found after injury, but is also seen in acute inflammatory disorders and in pigmented villonodular synovitis.

A single drop of fresh synovial fluid is placed on a glass slide and examined through the microscope. Blood cells are easily identified; abundant leucocytes may suggest infection. Crystals may be seen, though this usually requires a careful search; they are better characterized by polarized light microscopy (see Chapter 4).

Dry smears are prepared with heparinized fluid; more concentrated specimens can be obtained if the fluid is centrifuged. After suitable staining (Wright's and Gram's), the smear is examined for pus cells and organisms. Remember, though, that negative findings do not exclude infection.

Laboratory tests

If enough fluid is available, it is sent for full laboratory investigation (cells, biochemistry and bacteriological culture; see Table 1.3). A simultaneous blood specimen allows comparison of synovial and blood glucose concentration; a marked reduction of synovial glucose suggests infection.

A high white cell count (more than 10 000/mm^3) is usually indicative of infection, but a moderate leucocytosis is also seen in gout and other types of inflammatory arthritis.

Bacteriological culture and tests for antibiotic sensitivity are essential in any case of suspected infection.

BONE BIOPSY

Bone biopsy is often the crucial means of making a diagnosis or distinguishing between local conditions that closely resemble one another. Confusion is most likely to occur when the X-ray or MRI discloses an area of bone destruction that could be due to a compression fracture, a bone tumour or infection (e.g. a collapsed vertebral body). In other cases it is obvious that the lesion is a tumour – but what type of tumour? Benign or malignant? Primary or metastatic? Radical surgery should never be undertaken for a suspected neoplasm without first confirming the diagnosis histologically, no matter how 'typical' or 'obvious' the X-ray appearances may be.

In bone infection, the biopsy permits not only histological proof of acute inflammation but also bacteriological typing of the organism and tests for antibiotic sensitivity.

Table 1.3 Examination of synovial fluid

Suspected condition	Appearance	Viscosity	White cells	Crystals	Biochemistry	Bacteriology
Normal	Clear yellow	High	Few	–	As for plasma	–
Septic arthritis	Purulent	Low	+	–	Glucose low	+
Tuberculous arthritis	Turbid	Low	+	–	Glucose low	+
Rheumatoid arthritis	Cloudy	Low	++	–	–	–
Gout	Cloudy	Normal	++	Urate	–	–
Pseudogout	Cloudy	Normal	+	Pyrophosphate	–	–
Osteoarthritis	Clear yellow	High	Few	Often +	–	–

The investigation of metabolic bone disease sometimes calls for a tetracycline-labelled bone biopsy to show: (a) the type of abnormality (osteoporosis, osteomalacia, hyperparathyroidism), and (b) the severity of the disorder.

Open or closed?

Open biopsy, with exposure of the lesion and excision of a sizeable portion of the bone, seems preferable, but it has several drawbacks:

- It requires an operation, with the attendant risks of anaesthesia and infection.
- New tissue planes are opened up, predisposing to spread of infection or tumour.
- The biopsy incision may jeopardize subsequent wide excision of the lesion.
- The more inaccessible lesions (e.g. a tumour of the acetabular floor) can be reached only by dissecting widely through healthy tissue.

A carefully performed 'closed' biopsy, using a needle or trephine of appropriate size to ensure the removal of an adequate sample of tissue, is the procedure of choice except when the lesion cannot be accurately localized or when the tissue consistency is such that a sufficient sample cannot be obtained. Solid or semi-solid tissue is removed intact by the cutting needle or trephine; fluid material can be aspirated through the biopsy needle.

Precautions

- The biopsy site and approach should be carefully planned with the aid of X-rays or other imaging techniques.
- If there is any possibility of the lesion being malignant, the approach should be sited so that the wound and biopsy track can be excised if later radical surgery proves to be necessary.
- The procedure should be carried out in an operating theatre, under anaesthesia (local or general) and with full aseptic technique.
- For deep-seated lesions, fluoroscopic control of the needle insertion is essential.
- The appropriate size of biopsy needle or cutting trephine should be selected.
- A knowledge of the local anatomy and of the likely consistency of the lesion is important. Large blood vessels and nerves must be avoided; potentially vascular tumours may bleed profusely and the means to control haemorrhage should be readily to hand. More than one surgeon has set out to aspirate an 'abscess' only to plunge a wide-bore needle into an aneurysm!
- Clear instructions should be given to ensure that the tissue obtained at the biopsy is suitably processed. If infection is suspected, the material should go into a culture tube and be sent to the laboratory as soon as possible. A smear may also be useful. Whole tissue is transferred to a jar containing formalin, without damaging the specimen or losing any material. Aspirated blood should be allowed to clot and can then be preserved in formalin for later paraffin embedding and sectioning. Tissue thought to contain crystals should not be placed in formalin as this may destroy the crystals; it should be either kept unaltered for immediate examination or stored in saline.
- No matter how careful the biopsy, there is always the risk that the tissue will be too scanty or too unrepresentative for accurate diagnosis. Close consultation with the radiologist and pathologist beforehand will minimize this possibility. In the best hands, needle biopsy has an accuracy rate of over 95%.

DIAGNOSTIC ARTHROSCOPY

Arthroscopy is performed for both diagnostic and therapeutic reasons. Almost any joint can be reached but the procedure is most usefully employed in the knee, shoulder, wrist, ankle and hip. If the suspect lesion is amenable to surgery, it can often be dealt with at the same sitting without the need for an open operation. However, arthroscopy is an invasive procedure and its mastery requires skill and practice; it should not be used simply as an alternative to clinical examination and imaging.

Technique

The instrument is basically a rigid telescope fitted with fibreoptic illumination. Tube diameter ranges from about 2 mm (for small joints) to 4–5 mm (for the knee). It carries a lens system that gives a magnified image. The eyepiece allows direct viewing by the arthroscopist, but it is far more convenient to fit a small, sterilizable solid-state television camera which produces a picture of the joint interior on a television monitor.

The procedure is best carried out under general anaesthesia; this gives good muscle relaxation and permits manipulation and opening of the joint compartments. The joint is distended with fluid and the arthroscope is introduced percutaneously. Various instruments (probes, curettes and forceps) can be inserted through other skin portals; they are used to help expose the less accessible parts of the joint, or to obtain biopsies for further examination. Guided by the image on the monitor, the arthroscopist explores the joint in a systematic fashion, manipulating the arthroscope with one hand and the probe or forceps

with the other. At the end of the procedure the joint is washed out and the small skin wounds are sutured. The patient is usually able to return home later the same day.

Diagnosis

The knee is the most accessible joint. The appearance of the synovium and the articular surfaces usually allows differentiation between inflammatory and non-inflammatory, destructive and non-destructive lesions. Meniscal tears can be diagnosed and treated immediately by repair or removal of partially detached segments. Cruciate ligament deficiency, osteocartilaginous fractures, cartilaginous loose bodies and synovial 'tumours' are also readily visualized.

Arthroscopy of the shoulder is more difficult, but the articular surfaces and glenoid labrum can be adequately explored. Rotator cuff lesions can often be diagnosed and treated at the same time. Arthroscopy of the wrist is useful for diagnosing torn triangular fibrocartilage and interosseous ligament ruptures.

Arthroscopy of the hip is becoming more common and is proving to be useful in the diagnosis of unexplained hip pain. Labral tears, synovial lesions, loose bodies and articular cartilage damage (all of which are difficult to detect by conventional imaging techniques) have been diagnosed with a reported accuracy rate of over 50%.

Complications

Diagnostic arthroscopy is safe but not entirely free of complications, the commonest of which are haemarthosis, thrombophlebitis, infection and joint stiffness (particularly contracture of the anterior capsule). There is also a significant incidence of algodystrophy following arthroscopy.

FURTHER READING

Apley AG, Solomon L. *Physical Examination in Orthopaedics.* Oxford: Butterworth Heinemann, 1997.

Taylor & Francis
Taylor & Francis Group
http://taylorandfrancis.com

Infection

2

Enrique Gómez-Barrena

Infection – as distinct from mere residence of microorganisms – is a condition in which pathogenic microorganisms multiply and spread within the body tissues. Microorganisms may reach the musculoskeletal tissues by:

- *direct introduction* through the skin (a pinprick, an injection, a stab wound, a laceration, an open fracture or an operation, particularly when biomaterials are implanted),
- *direct spread from a contiguous focus of infection*, or
- *indirect spread via the bloodstream* from a distant site such as the nose or mouth, the respiratory tract, the bowel or the genitourinary tract.

Depending on the type of invader, the site of infection and the host response, the result may be a pyogenic osteomyelitis, a septic arthritis, a chronic granulomatous reaction (classically seen in tuberculosis of either bone or joint), or an indolent response to a less aggressive organism (as in low-grade periprosthetic infections) or to an unusual organism (e.g. a fungal infection). Soft-tissue infections range from superficial wound sepsis to widespread cellulitis and life-threatening necrotizing fasciitis. Parasitic lesions such as hydatid disease also are considered in this chapter, although these are infestations rather than infections.

Clinical aspects of infection will be particularly developed in this chapter. The team approach, including microbiologists, infectious disease and internal medicine doctors, is certainly the basis for success in infection. However, many cases of musculoskeletal infection need the leadership of the orthopaedic surgeon to ensure timely diagnosis and treatment of the patient with musculoskeletal symptoms and signs that suggest infection.

GENERAL ASPECTS OF INFECTION

Infection usually gives rise to an acute or chronic *inflammatory reaction*, which is the body's way of combating the invaders by destroying them, or at least immobilizing and confining them to a restricted area. The classical signs of inflammation are frequently present (*redness*, *swelling*, *heat*, *pain* and *loss of function*) and offer clinical clues about the infection and the patient's reaction.

Bone infection differs from soft-tissue infection since bone consists of a collection of rigid compartments. Bone is thus more susceptible than soft tissues to vascular damage and cell death due to pressure in acute inflammation. Unless it is rapidly suppressed, bone infection will inevitably lead to necrosis. *Osteomyelitis* is infection of bone and frequently seeds in trabecular areas affecting both bone and bone marrow. Soft-tissue infection depends on the main affected tissue, but of special interest to the orthopaedic surgeon is joint infection or *infectious arthritis*, whether *septic arthritis* or *granulomatous arthritis*. All these forms of infection will be addressed below.

Host susceptibility to infection is increased by (a) *local factors* such as trauma, scar tissue, poor circulation, diminished sensibility, chronic bone or joint disease and the presence of foreign bodies including implants, as well as (b) *systemic factors* such as malnutrition, general illness, debility, diabetes, rheumatoid disease, corticosteroid administration and all forms of immunosuppression, either acquired or induced.

BOX 2.1 FACTORS PREDISPOSING TO BONE INFECTION

Malnutrition and general debility
Diabetes mellitus
Corticosteroid administration
Immune deficiency
Immunosuppressive drugs
Venous stasis in the limb
Peripheral vascular disease
Loss of sensibility
Iatrogenic invasive measures
Trauma

Resistance is also diminished in the very young and the very old.

Bacterial colonization and resistance to antibiotics is enhanced by the ability of certain microorganisms (including *Staphylococcus*) to adhere to avascular bone surfaces and foreign implants, protected from both host defences and antibiotics by a protein–polysaccharide slime (*glycocalyx* or *biofilm*). Biofilm formation aids the development of a complex bacterial community that protects microorganisms adherent to biomaterials. Biofilm maturation with microorganism release further expands this colonization, and eradication of biofilm-forming microorganisms becomes impossible without implant removal or exchange. Thus, bacterial adherence to biomaterials and biofilm formation are crucial aspects to consider when treating musculoskeletal infections in the presence of implants.

Acute pyogenic bone infections are characterized by the formation of pus – a concentrate of defunct leucocytes, dead and dying bacteria and tissue debris – which is often localized in an abscess. Pressure builds up within the abscess and infection may then extend into a contiguous joint or through the cortex and along adjacent tissue planes. It may also spread further afield via lymphatics (causing lymphangitis and lymphadenopathy) or via the bloodstream (bacteraemia and septicaemia). An accompanying systemic reaction varies from a vague feeling of lassitude with mild pyrexia to severe illness, fever, toxaemia and shock. The generalized effects are due to the release of bacterial enzymes and endotoxins as well as cellular breakdown products from the host tissues.

Chronic pyogenic infection may follow unresolved acute infection and is characterized by persistence of the infecting organism (or, more frequently, multiple microorganisms) in pockets of necrotic tissue. Purulent material accumulates and may be discharged through sinuses at the skin or a poorly healed wound. Factors which predispose to this outcome are the presence of damaged muscle, dead bone (*sequestrum*) or a foreign implant, diminished local blood supply and a weak host response. Resistance is likely to be depressed in the very young and the very old, in states of malnutrition or immunosuppression, and in certain diseases such as diabetes and leukaemia.

Chronic non-pyogenic infection may result from invasion by organisms that produce a cellular reaction leading to the formation of granulomas consisting largely of lymphocytes, modified macrophages and multinucleated giant cells; this type of granulomatous infection is seen most typically in tuberculosis. Systemic effects are less acute but may ultimately be very debilitating, with lymphadenopathy, splenomegaly and tissue wasting.

Treatment

1 Identify the infecting organism and administer effective antibiotic treatment or chemotherapy.
2 Provide analgesia and general supportive measures, including rest of the affected part or splintage of the affected joint.
3 Release pus as soon as it is detected.
4 Eradicate avascular and necrotic tissue.
5 Stabilize the bone if it has fractured and restore continuity if there is a gap in the bone.
6 Maintain or regain soft-tissue and skin cover.

If treated early with effective antibiotics, acute infections can usually be cured. Once there is pus and bone necrosis, operative drainage will be needed.

When treating patients with bone or joint infection, it is wise to maintain continuous collaboration with a specialist in microbiology.

ACUTE HAEMATOGENOUS OSTEOMYELITIS

Aetiology and pathogenesis

Acute haematogenous osteomyelitis is mainly a disease of children. When adults are affected, it is usually because their resistance is lowered. Trauma may determine the site of infection, possibly by causing a small haematoma or fluid collection in a bone, in patients with concurrent bacteraemia. The incidence of acute haematogenous osteomyelitis in Western European children is thought to have declined in recent years, probably a reflection of improving social conditions. Different studies from the United Kingdom confirm a low incidence (less than 1 case per 100 000) in most recent periods although it is almost certainly much higher among less affluent populations. Also, a decrease in surgical treatment of those cases has been identified, possibly related to earlier and more effective antibiotic treatment.

The causal organism in both adults and children is usually *Staphylococcus aureus* (found in over 70% of cases), and less often one of the other Gram-positive cocci, such as the Group A beta-haemolytic streptococcus (*Streptococcus pyogenes*) which is found in chronic skin infections, as well as Group B streptococcus (especially in newborn babies) or the alphahaemolytic diplococcus *S. pneumoniae*. In children between 1 and 4 years of age, the Gram-negative *Haemophilus influenzae* used to be a fairly common pathogen for osteomyelitis and septic arthritis, but the introduction of *H. influenzae* type B vaccination in the 1990s has been followed by a much reduced incidence of this infection in many countries. In recent years its place has been taken by the increasing presence of *Kingella kingae*, mainly following upper respiratory infection in young children. Other Gram-negative organisms (e.g. *Escherichia coli*, *Pseudomonas aeruginosa*, *Proteus mirabilis* and the anaerobic *Bacteroides fragilis*) occasionally cause acute bone infection.

Curiously, patients with sickle-cell disease are prone to infection by *Salmonella typhi*. Anaerobic organisms (particularly *Peptococcus magnus*) have been found in patients with osteomyelitis, usually as part of a mixed infection. Unusual organisms are more likely to be found in heroin addicts and as opportunistic pathogens in patients with compromised immune defence mechanisms.

The bloodstream is invaded, perhaps from a minor skin abrasion, treading on a sharp object, an injection point, a boil, a septic tooth or – in the newborn – from an infected umbilical cord. *In adults*, the source of infection may be a urethral catheter, an indwelling arterial line or a contaminated needle and syringe. *In children*, the infection usually starts in the vascular metaphysis of a long bone, most often in the proximal tibia or in the distal or proximal ends of the femur. Predilection for this site has traditionally been attributed to the peculiar arrangement of the blood vessels in that area: the non-anastomosing terminal branches of the nutrient artery twist back in hairpin loops before entering the large network of sinusoidal veins; the relative vascular stasis and consequent lowered oxygen tension are believed to favour bacterial colonization. The structure of the fine vessels in the hypertrophic zone of the physis may more easily allow bacteria to pass through and adhere to type 1 collagen in that area. *In infants*, in whom there are still anastomoses between metaphyseal and epiphyseal blood vessels, infection can also reach the epiphysis (Figure 2.1). *In adults*, haematogenous infection accounts for only about 20% of cases of osteomyelitis. *Staphylococcus aureus* is the commonest organism but *Pseudomonas aeruginosa* often appears in patients using intravenous drugs. Adults with diabetes and vascular disease, who are prone to soft-tissue infections of the foot, may develop contiguous bone infection involving a variety of organisms.

Pathology

Acute haematogenous osteomyelitis shows a characteristic progression marked by *inflammation*, *suppuration*, *bone necrosis*, *reactive new bone formation* and, ultimately, *resolution and healing* or else intractable *chronicity*. However, the pathological picture varies considerably, depending on the patient's age, the site of infection, the virulence of the organism and the host response.

ACUTE OSTEOMYELITIS IN CHILDREN

The 'classical' picture is seen in children between 2 and 6 years. The earliest change in the metaphysis is an acute inflammatory reaction with vascular congestion, exudation of fluid and infiltration by polymorphonuclear leucocytes. The intraosseous pressure rises rapidly, causing intense pain, obstruction to blood flow and intravascular thrombosis. Even at an early stage, the bone tissue is threatened by impending ischaemia and resorption due to a combination of phagocytic activity and the local accumulation of cytokines, growth factors, prostaglandin and bacterial enzymes. By the second or third day, pus forms within the bone and forces its way along the Volkmann canals to the surface where it produces a subperiosteal abscess. This is much more evident in children, because of the relatively loose attachment of the periosteum, than in adults. From the subperiosteal abscess, pus can spread along the shaft, to re-enter the bone at another level or burst into the surrounding soft tissues. The developing physis acts as a barrier to direct spread towards the epiphysis, but where the metaphysis is partly

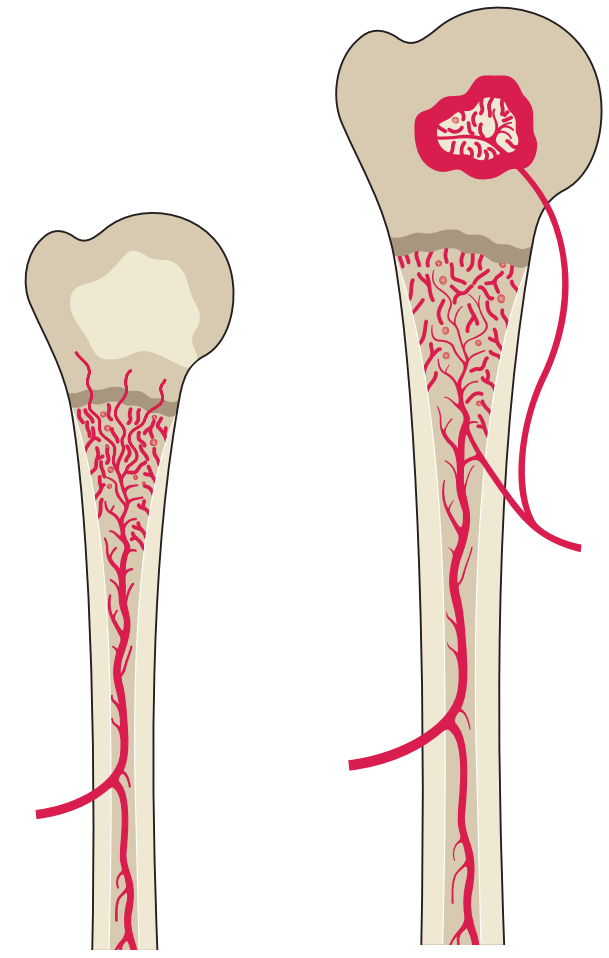


Figure 2.1 Epiphyseal and metaphyseal blood supply (a) In newborn infants some metaphyseal arterioles from the nutrient artery penetrate the physis and may carry infection directly from the metaphysis to the epiphysis. (b) In older children the physis acts as a barrier and the developing epiphysis receives a separate blood supply from the epiphyseal and periarticular blood vessels.

intracapsular (e.g. at the hip, shoulder or elbow) pus may discharge through the periosteum into the joint.

The rising intraosseous pressure, vascular stasis, small-vessel thrombosis and periosteal stripping increasingly compromise the blood supply; by the end of a week there is usually microscopic evidence of bone death. Bacterial toxins and leucocytic enzymes also may play their part in the advancing tissue destruction. With the gradual ingrowth of granulation tissue the boundary between living and devitalized bone becomes defined. Pieces of dead bone may separate as *sequestra* varying in size from mere spicules to large necrotic segments of the cortex in neglected cases.

Macrophages and lymphocytes arrive in increasing numbers and the debris is slowly removed by a combination of phagocytosis and osteoclastic resorption. A small focus in cancellous bone may be completely resorbed, leaving a tiny cavity, but a large cortical or cortico-cancellous sequestrum will remain entombed, inaccessible to either final destruction or repair.

Another feature of advancing acute osteomyelitis is new bone formation. Initially, the area around the infected zone is porotic (probably due to hyperaemia and osteoclastic activity) but if the pus is not released, either spontaneously or by surgical decompression, new bone starts forming on viable surfaces in the bone and from the deep layers of the stripped periosteum. This is typical of pyogenic infection, and fine streaks of subperiosteal new bone usually become apparent on X-ray by the end of the second week. With time, this new bone thickens to form a casement, or *involucrum*, enclosing the sequestrum and infected tissue. If the infection persists, pus and tiny sequestrated spicules of bone may discharge through perforations (*cloacae*) in the involucrum and track by sinuses to the skin surface (Figure 2.2).

If the infection is controlled and intraosseous pressure released at an early stage, this dire progress can be halted. The bone around the zone of infection becomes increasingly dense; this, together with the periosteal reaction, results in thickening of the bone. In some cases the normal anatomy may eventually be reconstituted; in others, though healing is sound, the bone is left permanently deformed.

If healing does not occur, a nidus of infection may remain locked inside the bone, causing pus and sometimes bone debris to be discharged intermittently through a persistent sinus (or several sinuses). The infection has now lapsed into *chronic osteomyelitis*, which may last for many years.

ACUTE OSTEOMYELITIS IN INFANTS

The early features of acute osteomyelitis in infants are much the same as those in older children. However, a significant difference, during the first year of life,

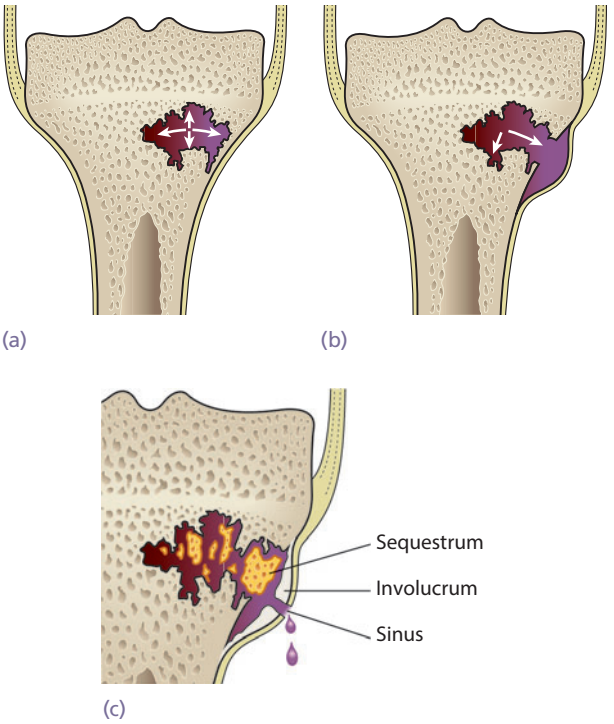


Figure 2.2 Acute osteomyelitis (a) Infection in the metaphysis may spread towards the surface, to form a subperiosteal abscess (b). Some of the bone may die and is encased in periosteal new bone as a sequestrum (c). The encasing involucrum is sometimes perforated by sinuses.

is the frequency with which the metaphyseal infection spreads to the epiphysis and from there into the adjacent joint. In the process, the physeal anlage may be irreparably damaged, further growth at that site is severely retarded and the joint will be permanently deformed. How this comes about is still argued over. It has long been held that, during the first 6–9 months of life, small metaphyseal vessels penetrate the physeal cartilage and this may permit the infection to spread into the cartilaginous epiphysis, although definitive proof of this mechanism has not been shown. Whatever the mechanism, what is indisputable is that osteomyelitis and septic arthritis often go together during infancy. Another feature in infants is an unusually exuberant periosteal reaction resulting in sometimes bizarre new bone formation along the diaphysis; fortunately, with longitudinal growth and remodelling, the diaphyseal anatomy is gradually restored.

ACUTE OSTEOMYELITIS IN ADULTS

Bone infection in the adult usually follows an open injury, an operation or spread from a contiguous focus of infection (e.g. a neuropathic ulcer or an infected diabetic foot) in over 70% of the cases. True haematogenous osteomyelitis is uncommon and, when it

does occur, it usually affects one of the vertebrae (e.g. following a pelvic infection), a metaphyseal region of a long bone or a small cuboidal bone. A vertebral infection may spread through the end plate and the intervertebral disc into an adjacent vertebral body. If a long bone is infected, the abscess is likely to spread within the medullary cavity, eroding the cortex and extending into the surrounding soft tissues. Periosteal new bone formation is less obvious than in childhood and the weakened cortex may fracture. If the bone end becomes involved, there is also a risk of the infection spreading into an adjacent joint.

Clinical features

Clinical features differ in the three described groups.

IN CHILDREN

The patient, usually a child over 4 years, presents with severe pain, malaise and a fever; in neglected cases, toxaemia may be marked. The parents will have noticed that he or she refuses to use one limb or to allow it to be handled or even touched. There may be a recent history of infection: a septic toe, a boil, a sore throat or a discharge from the ear. Typically the child looks ill and feverish; the pulse rate is likely to be over 100 and the temperature is raised. The limb is held still and there is acute tenderness near one of the larger joints (e.g. above or below the knee, in the popliteal fossa or in the groin). Even the gentlest manipulation is painful and joint movement is restricted ('pseudoparalysis'). Local redness, swelling, warmth and oedema are later signs and signify that pus has escaped from the interior of the bone. Lymphadenopathy is common but non-specific. *It is important to remember that all these features may be attenuated if antibiotics have been administered.*

IN INFANTS

In children under 1 year old, and especially in the newborn, the constitutional disturbance can be misleadingly mild; the baby simply fails to thrive and is drowsy but irritable. Suspicion should be aroused by a history of birth difficulties, umbilical artery catheterization or a site of infection (however mild) such as an inflamed intravenous infusion point or even a heel puncture. Metaphyseal tenderness and resistance to joint movement can signify either osteomyelitis or septic arthritis; indeed, both may be present, so the distinction hardly matters. Look for other sites – multiple infection is not uncommon, especially in babies who acquire the infection in hospital. Radionuclide bone scans may help to discover additional sites.

BOX 2.2 CARDINAL FEATURES OF ACUTE OSTEOMYELITIS IN CHILDREN

Pain
Fever
Refusal to bear weight
Elevated white blood cell count
Elevated ESR
Elevated CRP

IN ADULTS

A common site for haematogenous infection is the thoracolumbar spine. There may be a history of some urological procedure followed by a mild fever and backache. Local tenderness is not very marked and it may take weeks before X-ray signs appear; when they do appear, the diagnosis may still need to be confirmed by fine-needle aspiration and bacteriological culture. Other imaging may be required such as MRI, CT or SPECT-CT. In particular, deep abscess will need to be ruled out in case of suspicion, as surgical drainage may be required. Other bones are occasionally involved, especially if there is a background of diabetes, malnutrition, drug addiction, leukaemia, immunosuppressive therapy or debility. *In the very elderly, and in those with immune deficiency*, systemic features are mild and the diagnosis is easily missed.

Diagnostic imaging

PLAIN X-RAY

During the first week after the onset of symptoms, the plain radiograph shows no abnormality of the bone. Displacement of the fat planes signifies soft-tissue swelling, but this could as well be due to a haematoma or soft-tissue infection. By the second week there may be a faint extracortical outline due to periosteal new bone formation; this is the classic X-ray sign of early pyogenic osteomyelitis, but treatment should not be delayed while waiting for it to appear. Later, the periosteal thickening becomes more obvious and there is patchy rarefaction of the metaphysis; later still, the ragged features of bone destruction appear (Figure 2.3). An important late sign is the combination of regional osteoporosis with a localized segment of apparently increased density. Osteoporosis is a feature of metabolically active, and thus living, bone; the segment that fails to become osteoporotic is metabolically inactive and possibly dead.

ULTRASONOGRAPHY

Ultrasonography may detect a subperiosteal collection of fluid in the early stages of osteomyelitis, but it cannot distinguish between a haematoma and pus.

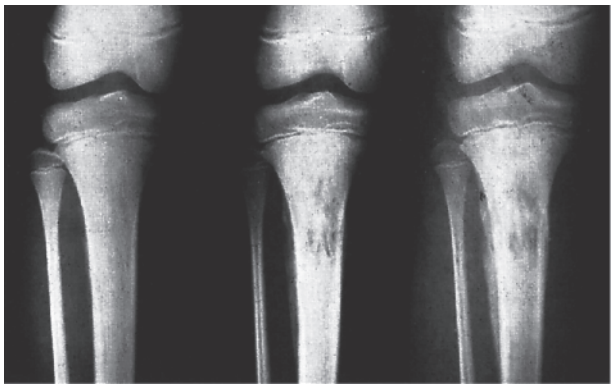


Figure 2.3 Acute osteomyelitis The first X-ray, 2 days after symptoms began, is normal – it always is; metaphyseal mottling and periosteal changes were not obvious until the second film, taken 14 days later; eventually much of the shaft was involved.

CT

Computed (or computerized) tomography offers the advantage of planar bone definition, including bone destruction and soft tissue mass, such as an abscess, within or surrounding bone. Disadvantages include high radiation dose. The excellent anatomical definition obtained from CT often justifies the higher radiation exposure.

RADIONUCLIDE SCANNING

Radioscintigraphy with ^{99}mTc-HDP reveals increased activity in both the perfusion phase and the bone phase. This is a highly sensitive investigation, even in the very early stages, but it has relatively low specificity and other inflammatory lesions can show similar changes. In doubtful cases, scanning with ^{67}Ga-citrate or ^{111}In-labelled leucocytes has been considered, but its use is decreasing in favour of other modalities.

SPECT/CT

As an alternative to radionuclide scanning, hybrid single-photon emission computed tomography/computer tomography (SPECT/CT) imaging is increasingly used in musculoskeletal infections. SPECT/CT imaging, compared with conventional planar study and SPECT alone, provides improved anatomic localization of infection and more accurate definition of its extent. Advantages of this modality include excellent differentiation between soft-tissue and bone infections, assessment of suspected infected sites with underlying structural bone alterations, and clear definition of infective foci within complex anatomical locations.

MAGNETIC RESONANCE IMAGING (MRI)

Magnetic resonance imaging can be helpful in cases of doubtful diagnosis, and particularly in suspected infection of the axial skeleton. It is also the best method of demonstrating bone marrow inflammation. It is extremely sensitive, even in the early phase of bone infection, and it can therefore assist in differentiating between soft-tissue infection and osteomyelitis. However, specificity is too low to exclude other local inflammatory lesions.

Laboratory investigations

The most certain way to confirm the clinical diagnosis is to aspirate pus or fluid from the metaphyseal subperiosteal abscess, the extraosseous soft tissues or an adjacent joint. This is best done using a 16- or 18-gauge trocar needle. Even if no pus is found, a smear of the aspirate is sent for detailed microbiological examination and tests for sensitivity to antibiotics. Immediate examination for cells and organisms through a simple Gram stain may help to identify the type of infection initially and assist with the early choice of antibiotic, but only until microbiological diagnosis through culture and antibiogram (the true etiological diagnosis to define specific treatment) is established. *Aspiration* will give a positive result in over 60% of cases that could be improved in case of open surgery by culture of tissue samples.

Blood cultures should be obtained if fever above 38 °C is detected, even though positive culture is obtained in less than half the cases of proven infection. The *C-reactive protein (CRP)* values are usually elevated within 12–24 hours and the *erythrocyte sedimentation rate (ESR)* within 24–48 hours after the onset of symptoms; both reactants (CRP+ESR) offer more information if simultaneously elevated. The *white blood cell (WBC) count* rises and the haemoglobin concentration may be diminished. *In the very young and the very old, these tests are less reliable and may show values within the range of normal.*

Anti-staphylococcal antibody titres may be raised. This test is useful in atypical cases where the diagnosis is in doubt.

Osteomyelitis in an unusual site or with an unusual organism should alert one to the possibility of heroin addiction, sickle-cell disease (*Salmonella* may be cultured from the faeces) or deficient host defence mechanisms including HIV infection.

Other tests such as IL-6 and alpha-defensin immunoassay are under evaluation, but their role is yet to be established.

Differential diagnosis

Cellulitis This is often mistaken for osteomyelitis. There is widespread superficial redness, with a clear demarcation between infected and normal skin, and lymphangitis. The source of skin infection may not be obvious and should be searched for (e.g. on the sole or between the toes). If doubt remains about the

diagnosis, MRI will help to distinguish between bone infection and soft-tissue infection. The organism is usually a staphylococcus or streptococcus. Mild cases will respond to high dosage oral antibiotics; severe cases need intravenous antibiotic treatment.

Acute suppurative arthritis Tenderness is diffuse, and movement at the joint is completely abolished by muscle spasm. In infants, the distinction between metaphyseal osteomyelitis and septic arthritis of the adjacent joint is somewhat theoretical, as both often coexist. A progressive rise in C-reactive protein values over 24–48 hours is considered suggestive of concurrent septic arthritis.

Streptococcal necrotizing myositis Group A beta-haemolytic streptococci (the same organisms which are responsible for the common 'sore throat') occasionally invade muscles and cause an acute myositis which, in its early stages, may be mistaken for cellulitis or osteomyelitis. Although the condition is rare, it should be kept well to the foreground in the differential diagnosis because it may rapidly spiral out of control towards muscle necrosis, septicaemia and death. Intense pain and board-like swelling of the limb in a patient with fever and a general feeling of illness are warning signs of a medical emergency. MRI will reveal muscle swelling and possibly signs of tissue breakdown. Immediate treatment with intravenous antibiotics is essential. Surgical debridement of necrotic tissue – and sometimes even amputation – may be needed to save a life.

Acute rheumatism The pain is less severe and it tends to flit from one joint to another. There may also be signs of carditis, rheumatic nodules or *erythema marginatum*.

Sickle-cell crisis The patient may present with features indistinguishable from those of acute osteomyelitis. In areas where *Salmonella* is endemic, it would be wise to treat such patients with suitable antibiotics until infection is definitely excluded.

Gaucher's disease 'Pseudo-osteitis' may occur with features closely resembling those of osteomyelitis. The diagnosis is made by finding other stigmata of the disease, especially enlargement of the spleen and liver.

Treatment

If osteomyelitis is suspected on clinical grounds, blood and fluid samples should be taken for laboratory investigation and then treatment started immediately without waiting for final confirmation of the diagnosis.

There are four important aspects to the management of the patient:

- appropriate antimicrobial therapy (first empirical, then specific)
- surgical drainage if required
- splintage and rest of the affected part
- supportive treatment for pain and dehydration.

ANTIBIOTICS

Blood and aspiration material are sent immediately for examination and culture, but the prompt intravenous administration of antibiotics is so vital that treatment should not await the result.

Initially, the choice of antibiotics is based on the findings from direct examination of the pus smear and the clinician's experience of local conditions – in other words, early empirical antibiotic administration, a 'best guess' at the most likely pathogen. *Staphylococcus aureus* is the most common at all ages, but treatment should provide cover also for other bacteria that are likely to be encountered in each age group; a more appropriate drug which is also capable of good bone penetration can be substituted, if necessary, once the infecting organism is identified and its antibiotic sensitivity is known. Factors such as the patient's age, general state of resistance, renal function, degree of toxaemia and previous history of allergy must be taken into account. The following classical recommendations are offered as a guide.

- *Neonates and infants up to 6 months of age*
  Initial antibiotic treatment should be effective against penicillin-resistant *Staphylococcus aureus*, Group B streptococcus and Gram-negative organisms. Drugs of choice are flucloxacillin plus a third-generation cephalosporin such as cefotaxime. Alternatively, effective empirical treatment can be provided by a combination of flucloxacillin (for penicillin-resistant staphylococci), benzylpenicillin (for Group B streptococci) and gentamicin (for Gram-negative organisms).
- *Children 6 months to 6 years of age*
  Empirical treatment in this age group should include cover against *Haemophilus influenzae*, unless it is known for certain that the child has had an anti-haemophilus vaccination. This is best provided by a combination of intravenous flucloxacillin and cefotaxime or cefuroxime.
- *Older children and previously fit adults*
  The vast majority in this group will have a staphylococcal infection and can be started on intravenous flucloxacillin and fusidic acid. Fusidic acid is preferred to benzylpenicillin partly because of the high prevalence of penicillin-resistant staphylococci and because it is particularly well concentrated in bone. However, for a known streptococcal infection benzylpenicillin is better. Patients who are allergic to penicillin should be treated with a polypeptide.

- *Elderly and previously unfit patients*
  In this group there is a greater than usual risk of Gram-negative infections, due to respiratory, gastrointestinal, or urinary disorders and the likelihood of the patient needing invasive procedures. The antibiotic of choice would be a combination of flucloxacillin and a second- or third-generation cephalosporin.
- *Patients with sickle-cell disease*
  These patients are prone to osteomyelitis, which may be caused by a staphylococcal infection but in many cases is due to *Salmonella* and/or other Gram-negative organisms. Chloramphenicol, which is effective against Gram-positive, Gram-negative and anaerobic organisms, used to be the preferred antibiotic, though there were always worries about the rare complication of aplastic anaemia. The current antibiotic of choice is a third-generation cephalosporin or a fluoroquinolone such as ciprofloxacin.
- *Heroin addicts and immunocompromised patients*
  Unusual infections (e.g. with *Pseudomonas aeruginosa*, *Proteus mirabilis* or anaerobic *Bacteroides* species) are likely in these patients. Infants with human immunodeficiency virus (HIV) infection may also have picked up other sexually transmitted organisms during birth. All patients with this type of background are therefore best treated empirically with a broad-spectrum antibiotic such as one of the third-generation cephalosporins or a fluoroquinolone preparation, depending on the results of sensitivity tests.
- *Patients considered to be at risk of methicillin-resistant Staphylococcus aureus (MRSA) infection*
  Patients admitted with acute haematogenous osteomyelitis and who have a previous history of MRSA infection, or any patient with a bone infection admitted to a hospital or a ward where MRSA is endemic, should be treated with intravenous vancomycin (or other glucopeptide such as teicoplanin) together with a third-generation cephalosporin. The usual programme is to administer the drugs intravenously (if necessary adjusting the choice of antibiotic once the results of antimicrobial sensitivity become available, and to the antibiotic trough and peak blood levels adjusted for the patient's kidney function and metabolism) until the patient's condition begins to improve and the CRP values return to normal levels – which usually takes 2–4 weeks depending on the virulence of the infection and the patient's general degree of fitness. By that time the most appropriate antibiotic would have been prescribed, on the basis of sensitivity tests; this can then be administered orally for another 3–6 weeks although, if bone destruction is marked, the period of treatment may have to be longer. While patients are on oral antibiotics, it is also important to track the serum antibiotic levels in order to ensure that the minimal inhibitory concentration (MIC) is maintained or exceeded. CRP, ESR and WBC values are also checked at regular intervals and treatment can be discontinued when these are seen to remain normal.

SURGICAL DRAINAGE

If antibiotics are given early (within the first 48 hours after the onset of symptoms), drainage is often unnecessary. However, if the clinical features do not improve within 36 hours of starting treatment, or even earlier, if there are signs of deep pus (swelling, oedema, fluctuation), and most certainly if pus is aspirated, the abscess should be drained by open surgery under general anaesthesia. If pus is found – and released – there is little to be gained by drilling into the medullary cavity. If there is no obvious abscess, it is reasonable to drill a few holes into the bone in various directions. There is no evidence that widespread drilling has any advantage and it may do more harm than good; if there is an extensive intramedullary abscess, drainage can be better achieved by cutting a small window in the cortex. The wound is closed without a drain and the splint (or traction) is reapplied. Once the signs of infection subside, movements are encouraged and the child is allowed to walk with the aid of crutches. Full weight-bearing is usually possible after 3–4 weeks.

At present, not more than one-third of patients with confirmed osteomyelitis are likely to need an operation and the percentage is decreasing; adults with vertebral infection seldom do.

SPLINTAGE

Some type of splintage is desirable, partly for comfort but also to prevent joint contractures. Simple skin traction may suffice and, if the hip is involved, this also helps to prevent dislocation. At other sites a plaster slab or half-cylinder may be used, but it should not obscure the affected area.

GENERAL SUPPORTIVE TREATMENT

The distressed child needs to be comforted and treated for pain. Analgesics should be given at repeated intervals without waiting for the patient to ask for them. Septicaemia and fever can cause severe dehydration and it may be necessary to give fluid intravenously.

Complications

A lethal outcome from septicaemia is nowadays extremely rare; with antibiotics the child nearly always recovers and the bone may return to normal. But morbidity and sequelae are common, especially if treatment is delayed or the organism is insensitive to the chosen antibiotic.

Epiphyseal damage and altered bone growth In neonates and infants whose epiphyses are still entirely cartilaginous, metaphyseal vessels penetrate the physis and may carry the infection into the epiphysis. If this happens, the physeal growth plate can be irrevocably damaged and the cartilaginous epiphysis may be destroyed, leading to arrest of growth and shortening of the bone. At the hip joint, the proximal end of the femur may be so badly damaged as to result in a pseudarthrosis.

Suppurative arthritis This may occur: (1) in very young infants, in whom the growth plate is not an impenetrable barrier; (2) where the metaphysis is intracapsular, as in the upper femur; or (3) from metastatic infection. In infants, it is so common as almost to be taken for granted, especially with osteomyelitis of the femoral neck. Ultrasound will help to demonstrate an effusion, but the definitive diagnosis is obtained by joint aspiration.

Metastatic infection This is sometimes seen – generally in infants – and may involve other bones, joints, serous cavities, the brain or lung. In some cases, the infection may be multifocal from the outset. Secondary infection sites are easily missed when attention is focused on one particular area; it is important to be alert to this complication and to repeatedly examine the child all over.

Pathological fracture Fracture is uncommon, but it may occur if treatment is delayed and the bone is weakened, either by erosion at the site of infection or by overzealous debridement.

Chronic osteomyelitis Despite improved methods of diagnosis and treatment, acute osteomyelitis sometimes fails to resolve. Weeks or months after the onset of acute infection, a sequestrum may appear in the follow-up X-ray and the patient may develop a chronic infection and a draining sinus. This may be related to late or inadequate treatment but is also seen in debilitated patients and in those with compromised defence mechanisms.

SUBACUTE HAEMATOGENOUS OSTEOMYELITIS

This condition is no longer rare, and in some countries the incidence is equal to that of acute osteomyelitis. Its relative mildness is presumably due to the organism being less virulent or the patient more resistant (or both). Its skeletal distribution is more variable than in acute osteomyelitis, but the distal femur and the proximal and distal tibia are the frequent sites. The anatomical classification (metaphyseal with or without cortical erosion, diaphyseal cortical or periosteal, epiphyseal, and vertebral) suggested by Roberts and colleagues in the 1980s is still helpful.

Pathology

Typically, there is a well-defined cavity in cancellous bone – usually in the tibial metaphysis – containing glairy seropurulent fluid (rarely pus). The cavity is lined by granulation tissue containing a mixture of acute and chronic inflammatory cells. The surrounding bone trabeculae are often thickened. The lesion sometimes encroaches on and erodes the bony cortex. Occasionally it appears in the epiphysis and, in adults, in one of the vertebral bodies.

Clinical features

The patient is usually a child or adolescent who has had pain near one of the larger joints for several weeks or even months. He or she may have a limp and often there is slight swelling, muscle wasting and local tenderness. The temperature is usually normal and there is little to suggest an infection. The WBC count and blood cultures usually show no abnormality but the ESR is sometimes elevated.

Imaging

The typical radiographic lesion is a circumscribed, round or oval radiolucent 'cavity' 1–2 cm in diameter. Most often it is seen in the tibial or femoral metaphysis, but it may occur in the epiphysis or in one of the cuboidal bones (e.g. the calcaneum). Sometimes the 'cavity' is surrounded by a halo of sclerosis (the classic *Brodie's abscess*); occasionally it is less well defined, extending into the diaphysis (Figure 2.4).

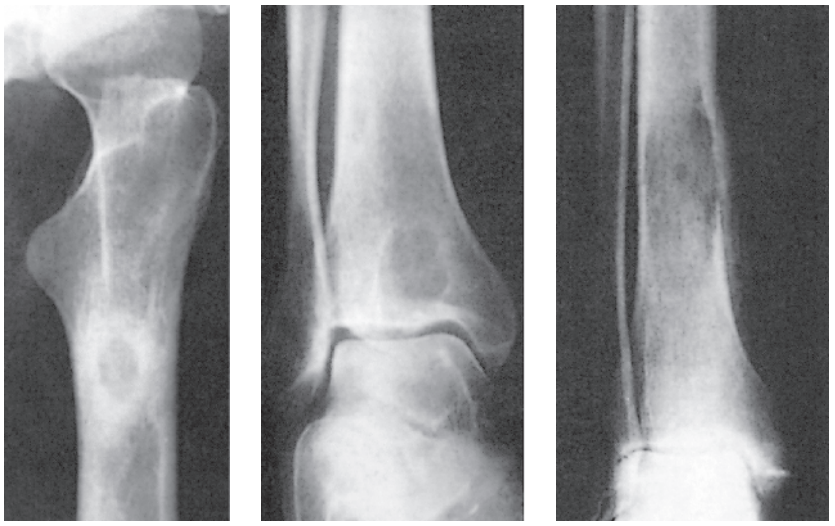
Metaphyseal lesions cause little or no periosteal reaction; diaphyseal lesions may be associated with periosteal new bone formation and marked cortical thickening. If the cortex is eroded, the lesion may be mistaken for a malignant tumour.

The radioisotope scan shows markedly increased activity.

Diagnosis

The clinical and X-ray appearances may resemble those of cystic tuberculosis, eosinophilic granuloma or osteoid osteoma; occasionally they mimic a malignant bone tumour such as Ewing's sarcoma. Epiphyseal lesions are easily mistaken for chondroblastoma. The diagnosis often remains in doubt until a biopsy is performed.

If fluid is encountered, it should be sent for bacteriological culture; this is positive in about half the cases and the organism is almost invariably *Staphylococcus aureus*.



(a) (b) (c)

Figure 2.4 Subacute osteomyelitis (a,b) The classic Brodie's abscess looks like a small walled-off cavity in the bone with little or no periosteal reaction. (c) Sometimes rarefaction is more diffuse and there may be cortical erosion and periosteal reaction.

Treatment

Treatment may be conservative if the diagnosis is not in doubt. Immobilization and antibiotics (flucloxacillin and fusidic acid) intravenously for 4 or 5 days and then orally for another 6 weeks usually result in healing, though this may take up to 12 months. If the diagnosis is in doubt, an open biopsy is needed and the lesion may be curetted at the same time. Curettage is also indicated if the X-ray shows that there is no healing after conservative treatment; this is always followed by a further course of antibiotics.

POST-TRAUMATIC OSTEOMYELITIS

Open fractures are always contaminated and are therefore prone to infection. The combination of tissue injury, vascular damage, oedema, haematoma, dead bone fragments and an open pathway to the atmosphere must invite bacterial invasion even if the wound is not contaminated with visible particulate dirt. *This is the most common cause of osteomyelitis in adults.*

Staphylococcus aureus is the usual pathogen, but other organisms such as *Escherichia coli*, *Proteus mirabilis* and *Pseudomonas aeruginosa* are sometimes involved. Occasionally, anaerobic organisms (clostridia, anaerobic streptococci or *Bacteroides*) appear in contaminated wounds.

Clinical features

The patient becomes feverish and develops pain and swelling over the fracture site; the wound is inflamed and there may be a seropurulent discharge. Blood tests reveal leucocytosis, increased CRP levels, and an elevated ESR; it should be remembered, though, that these inflammatory markers are non-specific and may be affected by tissue trauma.

X-ray appearances may be more difficult than usual to interpret because of bone fragmentation. *MRI* can be helpful in differentiating between bone and soft-tissue infection, but it is less reliable in distinguishing between long-standing infection and bone destruction due to trauma.

Microbiological investigation

If the wound is infected, a wound swab should be examined and cultured for organisms which can be tested for antibiotic sensitivity. Unfortunately, though, standard laboratory methods still yield negative results in about 20% of cases of overt infection. Routine wound swabs of open fracture wounds in the absence of infection is not recommended as cultured organisms are very unlikely to be the same as the organism causing any subsequent infection. Multiple tissue samples taken with clean, sterile instruments are preferred for microbiological investigations.

Treatment

The essence of treatment of open fractures is prophylaxis of infection: thorough cleansing and debridement of open fractures, the provision of drainage by leaving the wound open, immobilization of the fracture and antibiotics. In most cases a combination of flucloxacillin and benzylpenicillin (or sodium fusidate), given 6-hourly for 48 hours, will suffice. If the wound is clearly contaminated, it is wise also to give metronidazole for 4 or 5 days to control both aerobic and anaerobic organisms. Recent developments include the treatment of open fractures in one stage. This is a viable treatment option and can lead to good results if the soft tissue and bone debridement is meticulous and complete and adequate vascularized and tension free soft tissue closure can be obtained; this may require advance soft tissue procedures such as local or free flaps.

Pyogenic wound infection, once it has taken root, is difficult to eradicate. The presence of necrotic soft tissue and dead bone, together with a mixed bacterial flora, conspire against effective antibiotic control. Treatment requires soft tissue management and repeat debridement is required if there is evidence of inadequate debridement or infection.

Traditionally it was recommended that stable implants (fixation plates and intramedullary nails) should be left in place until the fracture had united, and this advice is still respected in recognition of the adage that even worse than an infected fracture is an *infected unstable* fracture. However, advances in external fixation techniques have meant that almost all fractures can, if necessary, be securely fixed by that method, with the added advantage that the wound remains accessible for dressings and superficial debridement. If these measures fail, the management is essentially that of chronic osteomyelitis.

CHRONIC OSTEOMYELITIS

This used to be the dreaded sequel to acute haematogenous osteomyelitis; nowadays, it more frequently follows an open fracture or an operation. The usual organisms (and with time there is always a mixed infection) are *Staphylococcus aureus*, *Escherichia coli*, *Streptococcus pyogenes*, *Proteus mirabilis* and *Pseudomonas aeruginosa*; in the presence of foreign implants *Staphylococcus epidermidis* (frequently coagulase negative staphylococcus), which is normally non-pathogenic, is the commonest of all.

Predisposing factors

Acute haematogenous osteomyelitis, if left untreated – and provided the patient does not succumb to septicaemia – will subside into a chronic bone infection which lingers indefinitely, perhaps with alternating 'flare-ups' and spells of apparent quiescence. The host defences are inevitably compromised by the presence of scar formation, dead and dying bone around the focus of infection, poor penetration of new blood vessels and non-collapsing cavities in which microbes can thrive. Bacteria covered in a protein–polysaccharide slime (*glycocalyx*) that protects them from both the host defences and antibiotics have the ability to adhere to inert surfaces such as bone sequestra and metal implants, where they multiply and colonize the area. There is also evidence that bacteria can survive inside osteoblasts and osteocytes and be released when the cells die.

These processes are evident in patients who have been inadequately treated ('too little too late'), but in any event certain patients are at greater risk than others: those who are very old or debilitated, those suffering from substance abuse and those with diabetes, peripheral vascular disease, skin infections, malnutrition, lupus erythematosus or any type of immune deficiency. The commonest of all predisposing factors is local trauma, such as an open fracture or a prolonged bone operation, especially if this involves the use of a foreign implant. Periprosthetic infection may evolve to chronic osteomyelitis and, due to its clinical relevance, will be addressed separately.

Pathology

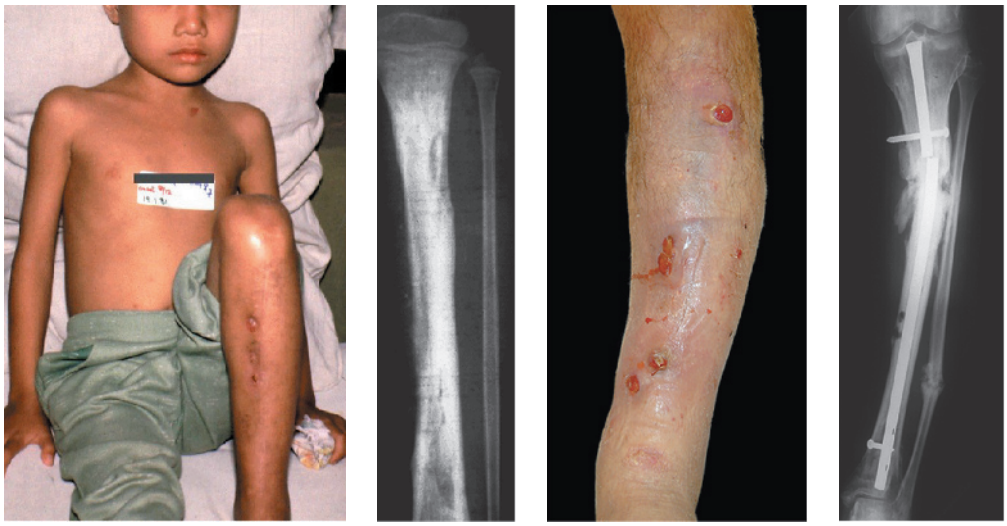
Bone is destroyed or devitalized, either in a discrete area around the focus of infection or more diffusely along the surface of an implant. Cavities containing pus and pieces of dead bone (sequestra) are surrounded by vascular tissue, and beyond that by areas of sclerosis – the result of chronic reactive new bone formation – which may take the form of a distinct bony sheath (involucrum). In the worst cases a sizeable length of the diaphysis may be devitalized and encased in a thick involucrum. Sequestra act as substrates for bacterial adhesion in much the same way as foreign implants, ensuring the persistence of infection until they are removed or discharged through perforations in the involucrum and sinuses that drain to the skin. A sinus may seal off for weeks or even months, giving the appearance of healing, only to reopen (or appear somewhere else) when the tissue tension rises. Bone destruction, and the increasingly brittle sclerosis, sometimes results in a pathological fracture. The histological picture is one of chronic inflammatory cell infiltration around areas of acellular bone or microscopic sequestra.

Clinical features

The patient presents because pain, pyrexia, redness and tenderness have recurred (a 'flare'), or with a discharging sinus. In long-standing cases, the tissues are thickened and often puckered or folded inwards where a scar or sinus adheres to the underlying bone. There may be a seropurulent discharge and excoriation of the surrounding skin. In post-traumatic osteomyelitis the bone may be deformed or ununited. See Figure 2.5.

Imaging

X-ray examination will usually show bone resorption – either as a patchy loss of density or as frank excavation around an implant – with thickening and sclerosis of the surrounding bone. However, there are marked variations: there may be no more than localized loss of trabeculation, or an area of osteoporosis, or periosteal thickening; sequestra show up as unnaturally dense fragments, in contrast to the surrounding osteopaenic bone; sometimes the bone is crudely thickened and misshapen, resembling a tumour. A *sinogram* may help to localize the site of infection.



(a) (b)

Figure 2.5 Chronic osteomyelitis
Chronic osteomyelitis may follow acute. The young boy (a) presented with draining sinuses at the site of a previous acute infection. The X-ray shows densely sclerotic bone. (b) In adults, chronic osteomyelitis is usually a sequel to open trauma or operation.

Radioisotope scintigraphy is sensitive but not specific. ^{99m}Tc-HDP scans show increased activity in both the perfusion phase and the bone phase. Scanning with ^{67}Ga-citrate or ^{111}In-labelled leucocytes is said to be more specific for osteomyelitis; such scans could be useful for showing up hidden foci of infection, although its low specificity has led to limited use.

CT and *MRI* are invaluable in planning operative treatment: together they will show the extent of bone destruction and reactive oedema, hidden abscesses and sequestra. *SPECT/CT* may provide advantages of sensitivity and local definition, and its use may increase in complex cases.

Investigations

During acute flares the CSR, ESR and WBC levels may be increased; these non-specific signs are helpful in assessing the progress of bone infection but they are not diagnostic.

Organisms cultured from discharging sinuses should be tested repeatedly for antibiotic sensitivity; with time, they often change their characteristics and become resistant to treatment. Note, however, that a superficial swab sample may not reflect the really persistent infection in the deeper tissues or may suffer from contamination; sampling from deeper tissues is crucial to understand the bone infection.

The most effective antibiotic treatment can be applied only if the pathogenic organism is identified and tested for sensitivity. Unfortunately, standard bacterial cultures still give negative results in about 20% of cases of overt infection. In recent years, more sophisticated molecular techniques have been developed, based on the amplification of bacterial DNA or RNA fragments (the polymerase chain reaction or PCR) and their subsequent identification by gel electrophoresis. However, although this has been shown to reveal unusual and otherwise undetected organisms in a significant percentage of cases, the technique is not widely available for routine testing.

A range of other investigations may also be needed to confirm or exclude suspected systemic disorders (such as diabetes) that could influence the outcome.

Table 2.1 Staging for adult chronic osteomyelitis

Lesion type	
Stage 1	Medullary
Stage 2	Superficial
Stage 3	Localized
Stage 4	Diffuse
Host category	
Type A	Normal
Type B	Compromised by local or systemic conditions
Type C	Severely compromised by local and systemic conditions

Staging of chronic osteomyelitis in long bones

'Staging' the condition helps in risk–benefit assessment and has some predictive value concerning the outcome of treatment. The system popularized by Cierny and colleagues in 2003 is based on both the local pathological anatomy and the host background (Table 2.1). The least serious, and most likely to benefit, are patients classified as Stage 1 or 2, Type A, i.e. those with localized infection and free of compromising disorders. Type B patients are somewhat compromised by a few local or systemic factors, but if the infection is localized and the bone still in continuity and stable (Stage 1–3) they have a reasonable chance of recovery. Type C patients are so severely

compromised that the prognosis is considered to be poor. If the lesion is also classified as Stage 4 (e.g. intractable diffuse infection in a non-united fracture), operative treatment may be contraindicated and the best option may be long-term palliative treatment known as suppression treatment. Occasionally one may have to advise amputation.

Treatment

ANTIBIOTICS

Chronic infection is seldom eradicated by antibiotics alone. Yet bactericidal drugs are important (a) to suppress the infection and prevent its spread to healthy bone and (b) to control acute flares. The choice of antibiotic depends on microbiological studies, but the drug must be capable of penetrating sclerotic bone and should be non-toxic with long-term use. Fusidic acid, clindamycin and the cephalosporins are good examples. Vancomycin and teicoplanin are effective in most cases of methicillin-resistant *Staphylococcus aureus* infection (MRSA).

Antibiotics are administered for 4–6 weeks (starting from the beginning of treatment or the last debridement) before considering operative treatment. During this time, serum antibiotic concentrations should be measured at regular intervals to ensure that they are kept at several times the minimal bactericidal concentration. *Continuous collaboration with a specialist in microbiology is important.* If surgical clearance fails, antibiotics should be continued for another 4 weeks before considering another attempt at full debridement.

LOCAL TREATMENT

A sinus may be painless and need dressing simply to protect the clothing. Colostomy paste can be used to stop excoriation of the skin. An acute abscess may need urgent incision and drainage, but this is only a temporary measure.

OPERATION

A waiting policy, punctuated by spells of bed rest and antibiotics to control flares, may have to be patiently endured until there is a clear indication for radical surgery: for *chronic haematogenous infections* this means intrusive symptoms, failure of adequate antibiotic treatment, and/or clear evidence of a sequestrum or dead bone; for *post-traumatic infections*, an intractable wound and/or an infected ununited fracture; for *postoperative infection*, similar criteria and evidence of bone erosion.

The presence of a *foreign implant* may prompt surgical intervention to remove the implant, whether in case of internal fixation (plates, screws and intramedullary nails that may be substituted by external fixation until infection control) or substitution, as discussed below. When undertaking operative treatment, collaboration with a plastic surgeon is strongly recommended.

Debridement At operation all infected soft tissue and dead or devitalized bone, as well as any infected implant, must be excised. The wound is inspected after 3 or 4 days and, if there are renewed signs of tissue death, the debridement may have to be repeated – several times if necessary. Antibiotic cover is continued for at least 4 weeks after the last debridement.

Dealing with the 'dead space' There are several ways of dealing with the resulting 'dead space'. *Porous antibiotic- impregnated beads* can be laid in the cavity and left for 2 or 3 weeks and then replaced with *cancellous bone grafts.* Bone grafts have also been used on their own; in the *Papineau technique* the entire cavity is packed with small cancellous chips (preferably autogenous) mixed with an antibiotic and a fibrin sealant. Where possible, the area is covered by adjacent muscle and the skin wound is sutured without tension. An alternative approach is to employ a *muscle flap transfer*: in suitable sites a large wad of muscle, with its blood supply intact, can be mobilized and laid into the cavity; the surface is later covered with a split-skin graft. In areas with too little adjacent muscle (e.g. the distal part of the leg), the same objective can be achieved by transferring a myocutaneous island flap on a long vascular pedicle. A free vascularized bone graft is considered to be a better option, provided the site is suitable and the appropriate facilities for microvascular surgery are available.

A different technique is the *Lautenbach approach*, involving radical excision of all avascular and infected tissue followed by closed irrigation and suction drainage, and an appropriate antibiotic solution in high concentration to allow the 'dead space' to be filled by vascular granulation tissue.

In refractory cases it may be possible to excise the infected and/or devitalized segment of bone completely and then close the gap by the *Ilizarov method* of 'transporting' a viable segment from the remaining diaphysis. This is especially useful if infection is associated with non-union after fracture.

Soft-tissue cover Last but not least, the bone must be adequately covered with skin. For small defects, split thickness skin grafts may suffice; for larger wounds local musculocutaneous flaps, or free vascularized flaps, are needed. Vacuum-assisted closure (VAC) may help when the deep infection is solved, not before.

Aftercare Success is difficult to measure; a minute focus of infection might escape the therapeutic onslaught, only to flare into full-blown osteomyelitis many years later. Prognosis should always be guarded; local trauma must be avoided and any recurrence of symptoms, however slight, should be taken seriously and investigated. The watchword is 'cautious optimism' – a 'probable cure' is better than no cure at all.

PERIPROSTHETIC INFECTION

Periprosthetic joint infection (PJI) is a specific type of infection related to joint replacement and a dreadful complication, potentially chronic, with significant clinical relevance for the affected patient, the treating surgeon and the health system. With an incidence of about 1–2% in hip arthroplasty, 2–3% in knee arthroplasty, 1–2% in the shoulder and even 3–5% in the elbow, the economic impact may represent a 5- to 10-fold cost increase compared to a primary arthroplasty. Patient risk factors include obesity, diabetes, rheumatoid arthritis and immunosuppressive treatments. Other risk factors include previous surgery, perioperative infection at a distant site, allogeneic blood transfusion, prolonged operative time and postoperative complications, including hematoma, superficial surgical site infection, wound drainage, and wound dehiscence.

A simple classification based on clinical manifestations differentiates early-onset PJI (if under 3 months after surgery), commonly initiated at operation through intraoperative contamination by relatively virulent microorganisms; delayed-onset PJI (after 3 months but before 12–24 months after surgery), by less virulent microorganisms but with same origin at operation; and late-onset PJI (more than 12–24 months after surgery), frequently due to haematogenous infection but occasionally due to very low-grade microorganisms with an extremely indolent infection initiated at the time of surgery. Tsukayama and colleagues popularized a classification that included early postoperative infection, haematogenous infection, and late chronic infection, adding a fourth type with positive intraoperative culture in a patient with revision for presumed aseptic failure, although some of these may not be true infections.

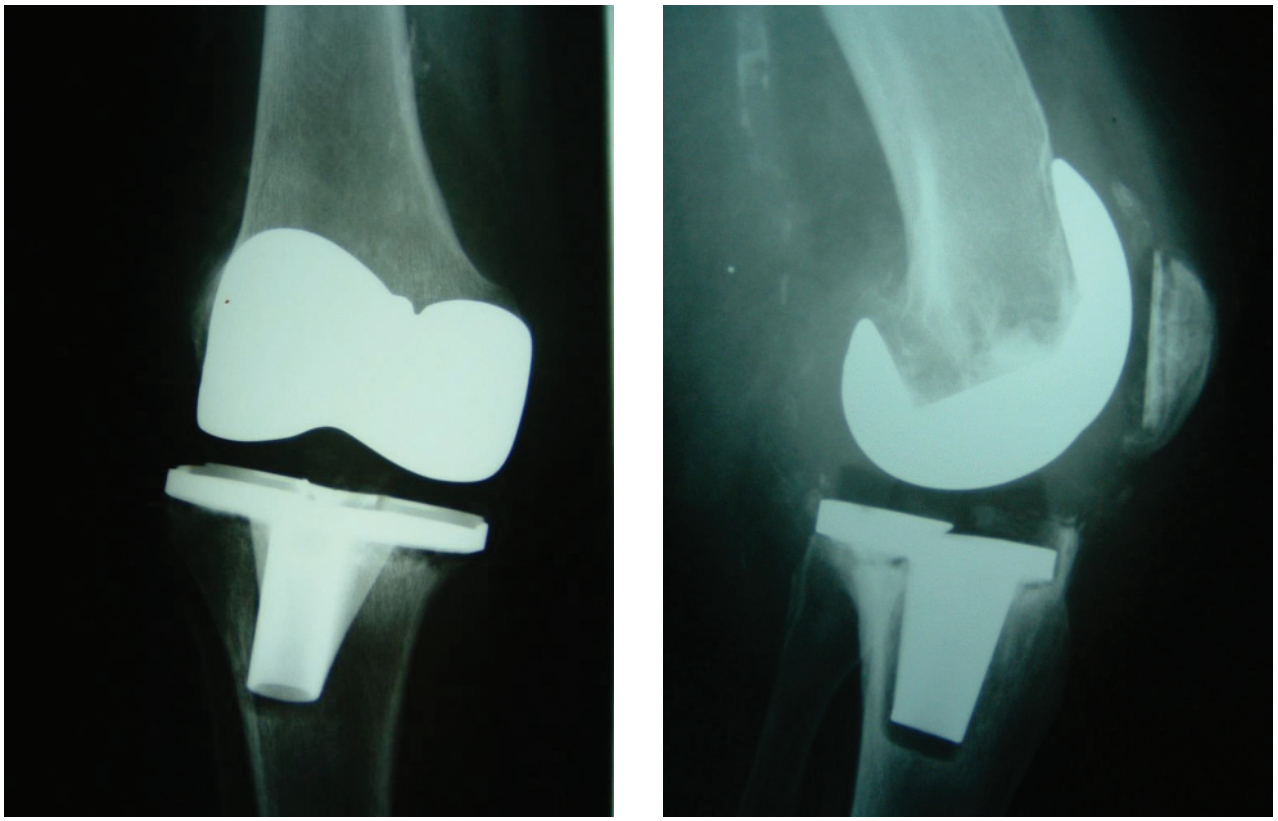
Adherence and biofilms Once in contact with the surface of the implant, microorganisms colonize the surface of the implant in competition with the host cells, in the so-called 'race for the surface'. After the initial adherence, the microorganisms colonize the implant through biofilm formation. Biofilms are complex communities of microorganisms embedded in an extracellular matrix formed on surfaces. From the attachment of microbial cells to a surface, the biofilm grows and matures until detachment and propagation, protecting microorganisms in a multicellular non-homogeneous structure where microbial cells communicate with one another (e.g. through quorum sensing) as in a multicellular organism protected from antibiotics and the host immune system. Clearing the biofilm requires surgical treatment, frequently with implant removal together with radical debridement of all infected tissues, followed by specific intravenous antibiotics.

The causative microorganism of PJI is most frequently *Staphylococcus aureus*, followed by coagulase-negative *Staphylococcus*. Together, these represent more than 50% of PJIs. *Streptococcus* species, *Enterococcus* species, aerobic Gram-negative bacilli, and some anaerobic (such as *Propionibacterium acnes* in the shoulder) account for 20–30%, while polymicrobial infections occur in 10–20%, the rest being culture-negative or other infrequent microorganisms. The increasing presence of multiresistant microorganisms requires carefully individualized antibiotic treatments.

The general diagnosis of PJI requires assessment of whether the joint is infected and, if so, determination of the causative microorganisms and their antimicrobial susceptibility. Thus, the diagnosis of PJI results from a combination of clinical findings, radiographic results (including early osteolysis and intraosseous abscesses, Figure 2.6), laboratory results (particularly CRP and ESR, WBC, but also IL-6 and procalcitonin) from peripheral blood and synovial fluid, microbiological data, intraoperative inspection, and histological evaluation of periprosthetic tissue. No single test offers sufficient accuracy alone. Clinically, definitive evidence of periprosthetic joint infection is obtained only when a sinus tract in communication with the prosthesis or an identical pathogen found in two separate periprosthetic tissue or fluid samples is confirmed. Other minor criteria also provide supportive (although not definitive) evidence, such as purulence surrounding the prosthesis, acute inflammation in periprosthetic histology, single virulent organism, elevated WBC, CRP and ESR.

Sonication of retrieved implants has been introduced to culture the dislodged biofilm and microorganisms from the surface of infected implants. Microorganisms were similarly obtained from all the different materials in the infected, retrieved prosthesis, proving that any remaining part of the implant may retain microorganisms, unless removed in revision surgery.

Treatment of periprosthetic joint infection usually requires both surgery and medical therapy, including prolonged antibiotic therapy after hospital discharge. The team approach, including surgeons, microbiologists, infectious disease physicians, nursing staff and other health professionals, is strictly required. Surgical treatment options oscillate from debridement with prosthesis retention (in early infections, particularly of haematogenous origin), one-stage arthroplasty exchange, two-stage arthroplasty exchange with or without antibiotic-loaded polymethylmethacrylate spacer, arthroplasty resection without reimplantation, or even suppression treatment consisting of long-term antibiotic treatment alone. Rarely, amputation may be required in case of vital risk for the patient.



(a) (b)

Figure 2.6 Periprosthetic joint infection Septic loosening surrounding the tibial stem in this case with PJI 3 years after total knee arthroplasty associated with immunodepression due to chemotherapy in the treatment of severe malignancy in (a) anteroposterior and (b) lateral radiographic views.

But besides the complex, multidisciplinary treatment of these infections, perioperative and postoperative prevention of PJI is a major aspect in the control of these severe entities.

GARRÉ'S SCLEROSING OSTEOMYELITIS

In 1893 Garré described a rare form of non-suppurative osteomyelitis which is characterized by marked sclerosis and cortical thickening. There is no abscess, only a diffuse enlargement of the bone at the affected site – usually the diaphysis of one of the tubular bones or the mandible. The patient is typically an adolescent or young adult with a long history of aching and slight swelling over the bone. Occasionally there are recurrent attacks of more acute pain accompanied by malaise and slight fever.

X-rays show increased bone density and cortical thickening; in some cases the marrow cavity is completely obliterated. There is no abscess cavity.

Diagnosis can be difficult. If a small segment of bone is involved, it may be mistaken for an osteoid osteoma. If there is marked periosteal layering of new bone, the lesion resembles a Ewing's sarcoma. The biopsy will disclose a low-grade inflammatory lesion with reactive bone formation. Microorganisms are seldom cultured but the condition is usually ascribed to a staphylococcal infection.

Treatment is by operation: the abnormal area is excised and the exposed surface thoroughly curetted. Bone grafts, bone transport or free bone transfer may be needed.

MULTIFOCAL NON-SUPPURATIVE OSTEOMYELITIS

This obscure disorder – it is not even certain that it is an infection – was first described in isolated cases in the 1960s and 1970s, and later in a more comprehensive report on 14 patients of mixed age and sex. It is now recognized that: (1) it is not as rare as initially suggested; (2) it comprises several different syndromes which have certain features in common; and (3) there is an association with chronic skin infection, especially pustular lesions of the palms

and soles (palmo-plantar pustulosis) and pustular psoriasis.

In children the condition usually takes the form of multifocal (often symmetrical), recurrent lesions in the long-bone metaphyses, clavicles and anterior ribcage; in adults the changes appear predominantly in the sterno-costo-clavicular complex and the vertebrae. In recent years the various syndromes have been drawn together under the convenient acronym SAPHO – standing for synovitis, acne, pustulosis, hyperostosis and osteitis.

Early osteolytic lesions show histological features suggesting a subacute inflammatory condition; in long-standing cases there may be bone thickening and round cell infiltration. The aetiology is unknown. Despite the local and systemic signs of inflammation, there is no purulent discharge and microorganisms have seldom been isolated. The two most characteristic clinical syndromes will be described.

Subacute recurrent multifocal osteomyelitis

This appears as an inflammatory bone disorder affecting mainly children and adolescents. Patients develop recurrent attacks of pain, swelling and tenderness around one or other of the long-bone metaphyses (usually the distal femur or the proximal or distal tibia), the medial ends of the clavicles or a vertebral segment. Over the course of several years multiple sites are affected, sometimes symmetrically and sometimes simultaneously; with each exacerbation, the child is slightly feverish and may have a raised ESR.

X-ray changes are characteristic. There are small lytic lesions in the metaphysis, usually closely adjacent to the physis. Some of these 'cavities' are surrounded by sclerosis; others show varying stages of healing. The clavicle may become markedly thickened. If the spine is affected, it may lead to collapse of a vertebral body. *Radioscintigraphy* shows increased activity around the lesions.

Biopsy of the lytic focus is likely to show the typical histological features of acute or subacute inflammation. In long-standing lesions there is a chronic inflammatory reaction with lymphocyte infiltration. Bacteriological cultures are almost invariably negative.

Treatment is entirely palliative; antibiotics have no effect on the disease. Although the condition may run a protracted course, the prognosis is good and the lesions eventually heal without complications.

Sterno-costo-clavicular hyperostosis

Patients are usually in their forties or fifties, and men are affected more often than women. Clinical and radiological changes are usually confined to the sternum and adjacent bones and the vertebral column. As with recurrent multifocal osteomyelitis, there is a curious association with cutaneous pustulosis. The usual complaint is of pain, swelling and tenderness around the sternoclavicular joints; sometimes there is also a slight fever and the ESR may be elevated. Patients with vertebral column involvement may develop back pain and stiffness.

X-rays show hyperostosis of the medial ends of the clavicles, the adjacent sternum and the anterior ends of the upper ribs, as well as ossification of the sternoclavicular and costoclavicular ligaments. Vertebral changes include sclerosis of individual vertebral bodies, ossification of the anterior longitudinal ligament, anterior intervertebral bridging, end-plate erosions, disc space narrowing and vertebral collapse. *Radioscintigraphy* shows increased activity around the sternoclavicular joints and affected vertebrae.

The condition usually runs a protracted course with recurrent 'flares'. There is no effective treatment but symptoms tend to diminish or disappear in the long term; however, the patient may be left with ankylosis of the affected joints.

INFANTILE CORTICAL HYPEROSTOSIS (CAFFEY'S DISEASE)

Infantile cortical hyperostosis, also known as Caffey's disease, is a rare disease of infants and young children. It usually starts during the first few months of life with painful swelling over the tubular bones and/or the mandible (see Figure 2.7). The child may be feverish and irritable, refusing to move the affected limb. Infection may be suspected but, apart from the swelling, there are no local signs of inflammation. The ESR, though, is usually elevated.

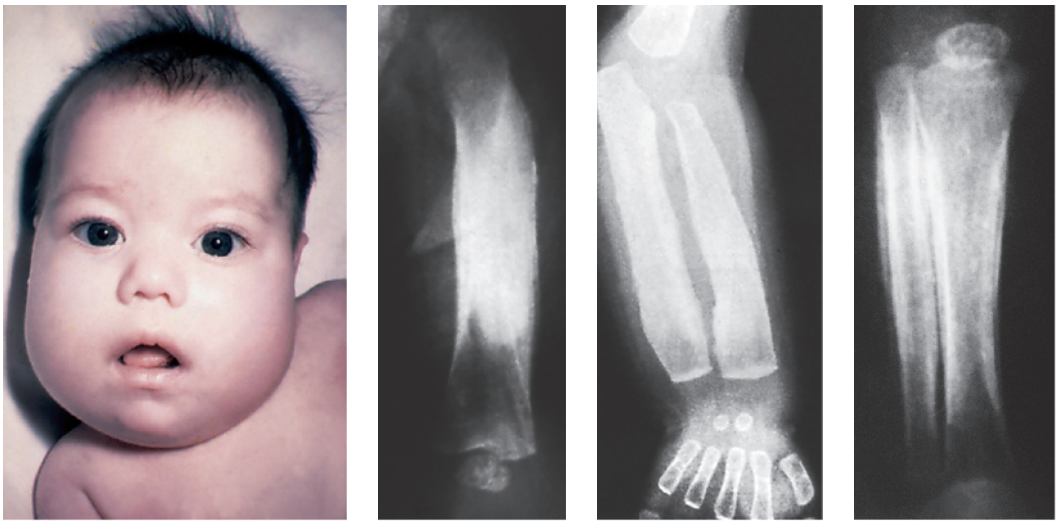
X-rays characteristically show periosteal new bone formation resulting in thickening of the affected bone.

After a few months the local features may resolve spontaneously, only to reappear somewhere else. Flat bones, such as the scapula and cranial vault, may also be affected.

Other causes of hyperostosis (osteomyelitis, scurvy) must be excluded. The cause of Caffey's disease is unknown but a virus infection has been suggested. Antibiotics are sometimes employed; it is doubtful whether they have any effect.

ACUTE SUPPURATIVE ARTHRITIS

A joint can become infected by: (1) direct invasion through a penetrating wound, intra-articular injection



(a) (b) (c) (d)

Figure 2.7 Caffey's disease This infant with Caffey's disease developed marked thickening of the mandible (a) and long bones. The lesions gradually cleared up, leaving little or no trace of their former ominous appearance.

or arthroscopy; (2) direct spread from an adjacent bone abscess; or (3) blood spread from a distant site. In infants it is often difficult to tell whether the infection started in the metaphyseal bone and spread to the joint or vice versa. In practice it hardly matters and in advanced cases it should be assumed that the entire joint and the adjacent bone ends are involved.

The causal organism is usually *Staphylococcus aureus*; however, in children between 1 and 4 years old, *Haemophilus influenzae* is an important pathogen unless they have been vaccinated against this organism. Occasionally other microbes, such as *Streptococcus*, *Escherichia coli* and *Proteus*, are encountered.

Predisposing conditions are rheumatoid arthritis, chronic debilitating disorders, intravenous drug abuse, immunosuppressive drug therapy and acquired immune deficiency syndrome (AIDS).

Pathology

The usual trigger is a haematogenous infection which settles in the synovial membrane; there is an acute inflammatory reaction with a serous or seropurulent exudate and an increase in synovial fluid. As pus appears in the joint, articular cartilage is eroded and destroyed, partly by bacterial enzymes and partly by proteolytic enzymes released from synovial cells, inflammatory cells and pus (Figure 2.8). In infants the entire epiphysis, which is still largely cartilaginous, may be severely damaged; in older children, vascular occlusion may lead to necrosis of the epiphyseal bone. In adults the effects are usually confined to the articular cartilage, but in late cases there may be extensive erosion due to synovial proliferation and ingrowth.

If the infection goes untreated, it will spread to the underlying bone or burst out of the joint to form abscesses and sinuses.

With healing there may be: (1) complete resolution and a return to normal; (2) partial loss of articular cartilage and fibrosis of the joint; (3) loss of articular cartilage and bony ankylosis; or (4) bone destruction and permanent deformity of the joint.

Clinical features

The clinical features differ somewhat according to the age of the patient.

In newborn infants the emphasis is on septicaemia rather than joint pain. The baby is irritable and refuses to feed; there is a rapid pulse and sometimes a fever. Infection is often suspected, but it could be anywhere! The joints should be carefully felt and moved to elicit the local signs of warmth, tenderness and resistance to movement. The umbilical cord should be examined for a source of infection. An inflamed intravenous infusion site should always excite suspicion. The baby's chest, spine and abdomen should be carefully examined to exclude other sites of infection. *Special care should be taken not to miss a concomitant osteomyelitis in an adjacent bone end.*

In children the usual features are acute pain in a single large joint (commonly the hip or the knee) and reluctance to move the limb ('pseudoparesis'). The child is ill, with a rapid pulse and a swinging fever. The overlying skin looks red and in a superficial joint swelling may be obvious. There is local warmth and marked tenderness. All movements are restricted, and often completely abolished, by pain and spasm. It is essential to look for a source of infection – a septic toe, a boil or a discharge from the ear.

In adults it is often a superficial joint (knee, wrist, a finger, ankle or toe) that is painful, swollen and inflamed. There is warmth and marked local tenderness, and movements are restricted. The patient

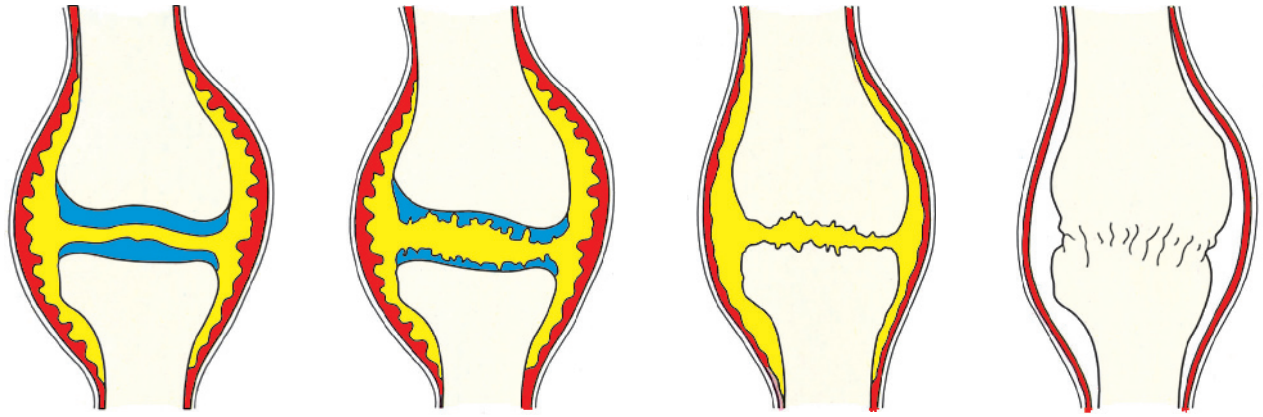


Figure 2.8 Acute suppurative arthritis – pathology In the early stage (a), there is an acute synovitis with a purulent joint effusion. (b) Soon the articular cartilage is attacked by bacterial and cellular enzymes. If the infection is not arrested, the cartilage may be completely destroyed (c). Healing then leads to bony ankylosis (d).

should be questioned and examined for evidence of gonococcal infection or drug abuse. Patients with rheumatoid arthritis, and especially those on corticosteroid treatment, may develop a 'silent' joint infection. Suspicion may be aroused by an unexplained deterioration in the patient's general condition; every joint should be carefully examined.

Imaging

Ultrasonography is the most reliable method for revealing a joint effusion in early cases. Both hips should be examined for comparison. Widening of the space between capsule and bone of more than 2 mm is indicative of an effusion, which may be echo-free (perhaps a transient synovitis) or positively echogenic (more likely septic arthritis).

X-ray examination is usually normal early on but signs to be watched for are soft-tissue swelling, loss of tissue planes, widening of the radiographic 'joint space' and slight subluxation (because of fluid in the joint) (Figure 2.9). With some infections there is sometimes gas in the joint. Narrowing and irregularity of the joint space are late features.

MRI and radionuclide imaging are helpful in diagnosing arthritis in obscure sites such as the sacroiliac and sternoclavicular joints.

Investigations

The white blood cell count, CRP and ESR are raised and blood culture may be positive. However, special investigations take time and it is much quicker (and usually more reliable) to aspirate the joint and examine the fluid. It may be frankly purulent but beware! – in early cases the fluid may look clear. A white cell count and Gram stain should be carried out immediately: the normal synovial fluid leucocyte count is under 300 per mL; it may be over 10 000 per mL in non-infective inflammatory disorders, but counts of over 50 000 per mL are highly suggestive of sepsis. Gram-positive cocci are probably *Staphylococcus aureus*; Gram-negative cocci are either *Haemophilus influenzae* or *Kingella kingae* (in children) or *Gonococcus* (in adults). Samples of fluid are also sent for full microbiological examination and tests for antibiotic sensitivity.

Differential diagnosis

Acute osteomyelitis In young children, osteomyelitis may be indistinguishable from septic arthritis; often one must assume that both are present.

Other types of infection *Psoas abscess* and local *infection of the pelvis* must be kept in mind. Systemic features will obviously be the same as those of septic arthritis.

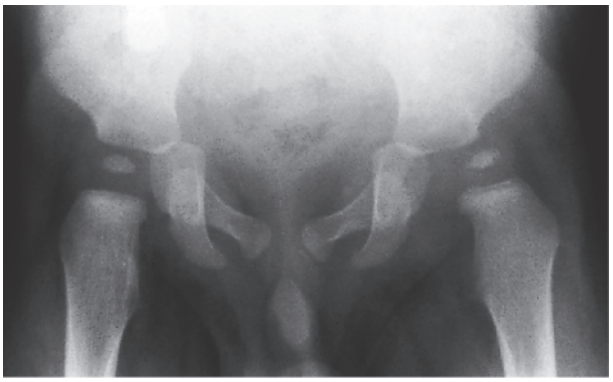
Trauma Traumatic synovitis or haemarthrosis may be associated with acute pain and swelling. A history of injury does not exclude infection. Diagnosis may remain in doubt until the joint is aspirated.

Irritable joint At the onset the joint is painful and lacks some movement, but the child is not really ill and there are no signs of infection. Ultrasonography may help to distinguish septic arthritis from transient synovitis.

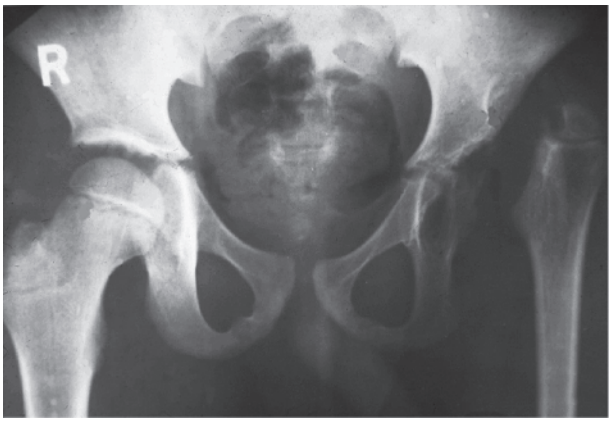
Haemophilic bleed An acute haemarthrosis closely resembles septic arthritis. The history is usually conclusive, but aspiration will resolve any doubt.

Rheumatic fever Typically the pain flits from joint to joint, but at the onset one joint may be misleadingly inflamed. However, there are no signs of septicaemia.

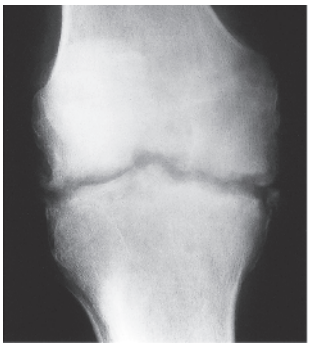
Juvenile rheumatoid arthritis This may start with pain and swelling of a single joint, but the onset is



(a)



(b)



(c)

Figure 2.9 Suppurative arthritis – X-ray (a) In this child the left hip is subluxated and the soft tissues are swollen. (b) If the infection persists untreated, the cartilaginous epiphysis may be entirely destroyed, leaving a permanent pseudarthrosis. (c) Septic arthritis in an adult knee joint.

usually more gradual and systemic symptoms less severe than in septic arthritis.

Sickle-cell disease The clinical picture may closely resemble that of septic arthritis – and indeed the bone nearby may actually be infected! – so this condition should always be excluded in communities where the disease is common.

Gaucher's disease In this rare condition acute joint pain and fever can occur without any organism being found ('pseudo-osteitis'). Because of the predisposition to true infection, antibiotics should be given.

Gout and pseudogout *In adults*, acute crystal-induced synovitis may closely resemble infection. On aspiration the joint fluid is often turbid, with a high white blood cell count; however, microscopic examination by polarized light will show the characteristic crystals.

Treatment

The first priority is to aspirate the joint and examine the fluid. Treatment is then started without further delay and follows the same lines as for acute osteomyelitis. Once the blood and tissue samples have been obtained, there is no need to wait for detailed results before giving antibiotics. If the aspirate looks purulent, the joint should be drained without waiting for laboratory results (see below).

DRAINAGE

Under anaesthesia the joint is opened through a small incision, drained and washed out with physiological saline. A small catheter is left in place and the wound is closed; suction–irrigation is continued for another 2 or 3 days. This is the safest policy and is certainly advisable (1) in very young infants, (2) when the hip is involved, and (3) if the aspirated pus is very thick. For the knee, arthroscopic debridement and copious irrigation may be equally effective. Older children with early septic arthritis (symptoms for less than 3 days) involving any joint except the hip can often be treated successfully by repeated closed aspiration of the joint; however, if there is no improvement within 48 hours, open drainage will be necessary.

ANTIBIOTICS

Antibiotic treatment follows the same guidelines as presented for acute haematogenous osteomyelitis. The initial choice of antibiotics is based on judgement of the most likely pathogens.

Neonates and infants up to the age of 6 months should be protected against staphylococcus and Gram-negative streptococci with one of the penicillinase-resistant penicillins (e.g. flucloxacillin) plus a third-generation cephalosporin.

Children from 6 months to puberty can be treated similarly. There is a risk of *Haemophilus* infection if they have not been immunized.

Older teenagers and adults can be started on flucloxacillin and fusidic acid. If the initial examination shows Gram-negative organisms a third-generation cephalosporin is added. More appropriate drugs can be substituted after full microbiological investigation. Antibiotics should be given intravenously for 4–7 days and then orally for another 3 weeks.

SPLINTAGE

The joint should be rested, and for neonates and infants this may mean light splintage; with hip infection, the

joint should be held abducted and 30 degrees flexed, on traction to prevent dislocation.

GENERAL SUPPORTIVE CARE

Analgesics are given for pain and intravenous fluids for dehydration.

AFTERCARE

Once the patient's general condition is satisfactory and the joint is no longer painful or warm, further damage is unlikely. If articular cartilage has been preserved, gentle and gradually increasing active movements are encouraged. If articular cartilage has been destroyed, the aim is to keep the joint immobile while ankylosis is awaited. Splintage in the optimum position is therefore continuously maintained, usually by plaster, until ankylosis is sound.

Complications

Infants under 6 months of age have the highest incidence of complications, most of which affect the hip. The most obvious risk factors are a delay in diagnosis and treatment (more than 4 days) and concomitant osteomyelitis of the proximal femur.

Subluxation and dislocation of the hip, or instability of the knee should be prevented by appropriate posturing or splintage.

Damage to the cartilaginous physis or the epiphysis in the growing child is the most serious complication. Sequelae include *retarded growth*, *partial or complete destruction of the epiphysis*, *deformity of the joint*, *epiphyseal osteonecrosis*, *acetabular dysplasia* and *pseudarthrosis of the hip.*

Articular cartilage erosion (chondrolysis) is seen in older patients and this may result in restricted movement or complete *ankylosis of the joint.*

GONOCOCCAL ARTHRITIS

Neisseria gonorrhoeae is the commonest cause of septic arthritis in sexually active adults, especially among poorer populations. Even in affluent communities the incidence of sexually transmitted diseases has increased (probably related to the increased use of non-barrier contraception) and with it the risk of gonococcal and syphilitic bone and joint diseases and their sequelae. The infection is acquired only by direct mucosal contact with an infected person – carrying a risk of greater than 50% after a single contact!

Clinical features

Two types of clinical disorder are recognized: (a) *disseminated gonococcal infection* – a triad of polyarthritis, tenosynovitis and dermatitis – and (b) *septic arthritis of a single joint* (usually the knee, ankle, shoulder, wrist or hand). Both syndromes may occur in the same patient. There may be a slight pyrexia and the ESR and WBC count will be raised. If the condition is suspected, the patient should be questioned about possible contacts during the previous days or weeks and they should be examined for other signs of genitourinary infection (e.g. a urethral discharge or cervicitis).

Joint aspiration may reveal a high white blood cell count and typical Gram-negative organisms, but bacteriological investigations are often disappointing. Samples should also be taken from the various mucosal surfaces and tests should be performed for other sexually transmitted infections.

Treatment

Treatment is similar to that of other types of pyogenic arthritis. Patients will usually respond quite quickly to a third-generation cephalosporin given intravenously or intramuscularly. However, bear in mind that many patients with gonococcal infection also have chlamydial infection, which is resistant to cephalosporins; both are sensitive to quinolone antibiotics such as ciprofloxacin and ofloxacin. If the organism is found to be sensitive to penicillin (and the patient is not allergic), treatment with ampicillin or amoxicillin and clavulanic acid is also effective.

SEPTIC ARTHRITIS AND HIV-1 INFECTION

Septic arthritis has been encountered quite frequently in HIV-positive intravenous drug users, HIV-positive haemophiliacs and other patients with AIDS. The usual organisms are *Staphylococcus aureus* and *Streptococcus*; however, opportunistic infection by unusual organisms is not uncommon.

The patient may present with an acutely painful, inflamed joint and marked systemic features of bacteraemia or septicaemia. In some cases the infection is confined to a single, unusual site such as the sacroiliac joint; in others several joints may be affected simultaneously. Opportunistic infection by unusual organisms may produce a more indolent clinical picture.

Treatment follows the general principles outlined before. Patients with staphylococcal and streptococcal infections usually respond well to antibiotic treatment and joint drainage; opportunistic infections may be more difficult to control.

SPIROCHAETAL INFECTIONS

Two conditions which are likely to be encountered by the orthopaedic surgeon are dealt with here: *syphilis*

and *yaws*. *Lyme disease*, which also originates with a spirochaetal infection, is better regarded as due to a systemic autoimmune response and is discussed in Chapter 3.

Syphilis

Syphilis is caused by the spirochaete *Treponema pallidum*, generally acquired during sexual activity by direct contact with infectious lesions of the skin or mucous membranes. The infection spreads to the regional lymph nodes and thence to the bloodstream. The organism can also cross the placental barrier and enter the fetal blood stream directly during the latter half of pregnancy, giving rise to congenital syphilis.

In acquired syphilis a *primary* ulcerous lesion, or *chancre*, appears at the site of inoculation about a month after initial infection. This usually heals without treatment but, a month or more after that, the disease enters a *secondary phase* characterized by the appearance of a maculopapular rash and bone and joint changes due to periostitis, osteitis and osteochondritis. After a variable length of time, this phase is followed by a *latent period* which may continue for many years. The term is somewhat deceptive because in about half the cases pathological lesions continue to appear in various organs and 10–30 years later the patient may present again with tertiary syphilis, which takes various forms including the appearance of large granulomatous gummata in bones and joints and neuropathic disorders in which the loss of sensibility gives rise to joint breakdown (*Charcot joints*).

In congenital syphilis, the primary infection may be so severe that the fetus is either stillborn or the infant dies shortly after birth. The ones who survive manifest pathological changes similar to those described above, though with modified clinical appearances and a contracted timescale. See Figure 2.10.

Clinical features of acquired syphilis

Early features The patient usually presents with pain, swelling and tenderness of the bones, especially those with little soft-tissue covering, such as the frontal bones of the skull, the anterior surface of the tibia, the sternum and the ribs. *X-rays* may show typical features of *periostitis* and *thickening of the cortex* in these bones, as well as others that are not necessarily symptomatic. *Osteitis* and *septic arthritis* are less common. Occasionally these patients develop polyarthralgia or polyarthritis. Enquiry may reveal a history of sexually transmitted disease.

Late features The typical late feature, which may appear only after many years, is the syphilitic *gumma*, a dense granulomatous lesion associated with local bone resorption and adjacent areas of sclerosis. Sometimes this results in a pathological fracture. *X-rays* may show thick periosteal new bone formation at other sites, especially the tibia.

The other well-recognized feature of tertiary syphilis is a neuropathic arthropathy due to loss of sensibility in the joint – most characteristically the knee.

Other neurological disorders, the early signs of which may only be discovered on careful examination, are tabes dorsalis and 'general paralysis of the insane' (GPI). With modern treatment, these late sequelae have become rare.

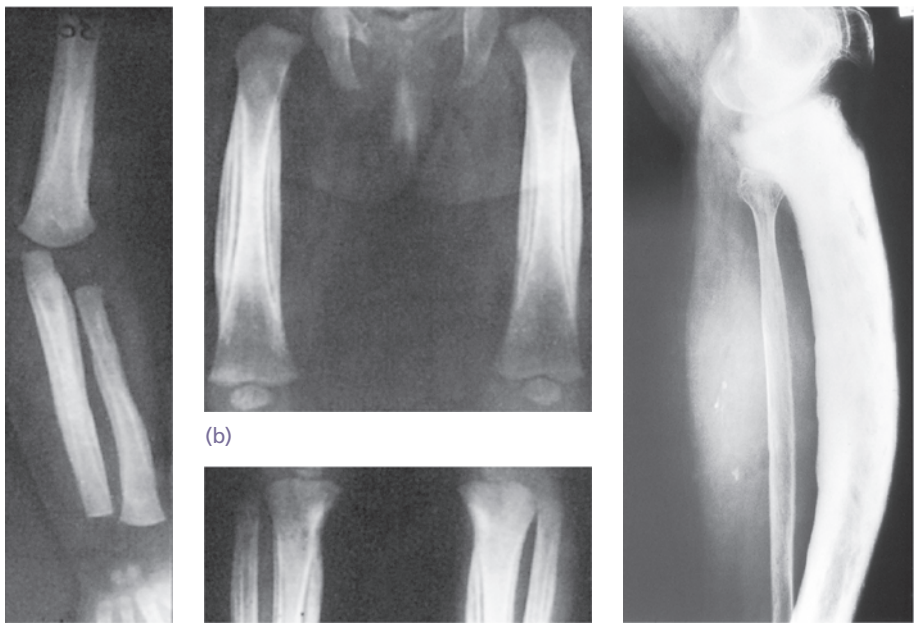


Figure 2.10 Syphilis (a–c) Congenital syphilis, with diffuse periostitis of many bones. (d) Acquired syphilitic periostitis of the tibia.

Clinical features of congenital syphilis

Early congenital syphilis Although the infection is present at birth, bone changes do not usually appear until several weeks afterwards. The baby is sick and irritable and examination may show skin lesions, hepatosplenomegaly and anaemia. Serological tests are usually positive in both mother and child.

The first signs of skeletal involvement may be joint swelling and 'pseudoparalysis' – the child refuses to move a painful limb. Several sites may be involved, often symmetrically, with slight swelling and tenderness at the ends or along the shafts of the tubular bones. The characteristic *X-ray changes* are of two kinds: *osteochondritis* (*'metaphysitis'*) – trabecular erosion in the juxta-epiphyseal regions of tubular bones showing first as a lucent band near the physis and later as frank bone destruction which may result in epiphyseal separation; and, less frequently, *periostitis* – diffuse periosteal new bone formation along the diaphysis, usually of mild degree but sometimes producing an 'onion-peel' effect. The condition must be distinguished from scurvy (rare in the first 6 months of life), multifocal osteomyelitis, the battered baby syndrome and Caffey's disease (see above).

Late congenital syphilis Bone lesions in older children and adolescents resemble those of acquired syphilis and some features occurring 10 or 15 years after birth may be manifestations of tertiary disease, the result of gumma formation and endarteritis. Gummata appear either as discrete, punched-out radiolucent areas in the medulla or as more extensive destructive lesions in the cortex. The surrounding bone is thick and sclerotic. Sometimes the predominant feature is dense endosteal and periosteal new bone formation affecting almost the entire bone (the classic '*sabre tibia*').

Other abnormalities which have come to be regarded as 'classic' features in older children are dental malformations ('Hutchinson's teeth'), erosion of the nasal bones, thickening and expansion of the finger phalanges (dactylitis) and painless effusions in the knees or elbows ('Clutton's joints').

Treatment

Early lesions will usually respond to intramuscular injections of benzylpenicillin given weekly for 3 or 4 doses. Late lesions will require high-dosage intravenous penicillin for a week or 10 days, but some forms of tertiary syphilis will not respond at all. An alternative would be treatment with one of the third-generation cephalosporins.

Yaws

Yaws is a non-venereal spirochaetal infection caused by *Treponema pertenue*. It is seen mainly in the poorer tropical parts of Africa, Asia and South America. Though considered – at least in Europe – to be a 'rare' disease, several thousand cases a year are reported in Indonesia.

The infection is contracted by skin-to-skin contact. A knobbly ulcer covered by a scab (the *primary* or '*mother*' *yaw*), usually develops on the face, hands or feet. Secondary skin lesions appear 1–4 months later and successive lesions may go on to pustular ulceration; as each one heals it leaves a pale tell-tale scar. This *secondary stage* is followed by a long latent period, merging into a *tertiary stage* during which skeletal changes similar to those of syphilis develop – periosteal new bone formation, cortical destruction and osteochondritis.

Clinical features

Children under 10 years old are the usual victims. In areas where the disease is endemic, the typical skin lesions and an associated lymphadenopathy are quickly recognized. Elsewhere, further investigations may be called for – serological tests and dark-field examination of scrapings from one of the skin lesions.

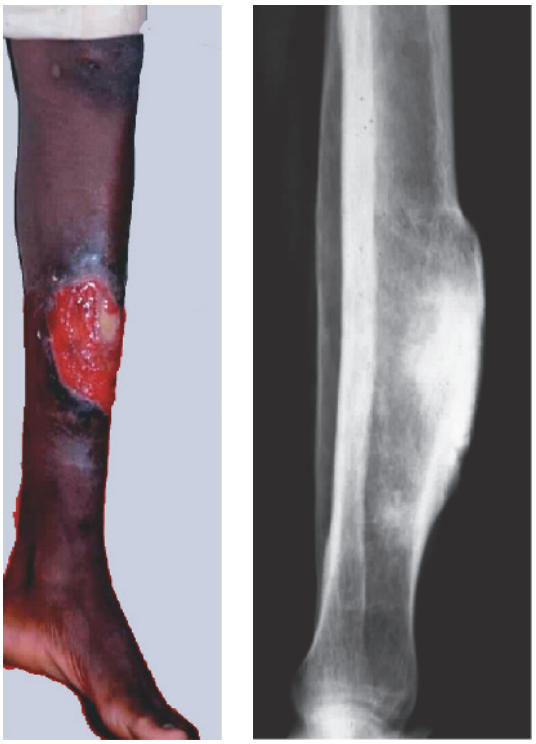
At a later stage deformities and bone tenderness may become apparent. *X-rays* show features such as cortical erosion, joint destruction and periosteal new bone formation; occasionally thickening of a long bone may be so marked as to resemble the 'sabre tibia' of late congenital syphilis.

Treatment

Treatment with benzylpenicillin, preferably given by intramuscular injection, is effective. For those who are hypersensitive to penicillin, erythromycin is a satisfactory alternative.

TROPICAL ULCER

Tropical ulcer, though the name sounds vague and non-specific, is a distinct entity that is seen frequently in tropical and subtropical regions, particularly in parts of Africa, where people walk bare-legged through rough terrain or long grass. It almost always occurs on the leg and men make up the majority of patients. The initial lesion is a small split in the skin (a cut, thorn-scratch, insect bite or other minor abrasion), which is then contaminated with all kinds of dirt or stagnant water. The most likely infecting organisms are *Fusiformis fusiformis* and *Borrelia vincentii* (both common in faeces). This results in an indolent ulcer which defies most forms of topical treatment (and certainly traditional remedies native to those parts of the world)



(a) (b)

Figure 2.11 Tropical ulcer (a) What started as a small ulcer has turned into a large spreading lesion. (b) The X-ray shows the typical marked periosteal reaction in the underlying bone.

(Figure 2.11). The ulcer may eventually bore its way into the soft tissues and the underlying bone; occasionally, after many years, it gives rise to a locally invasive squamous-cell carcinoma.

Clinical features

What starts as a small inflamed scratch or cut develops over a few days into a large pustule. By the time the patient attends for medical treatment, the pustule has usually ruptured, leaving a foul-smelling, discharging ulcer with hard rolled edges on the leg, the ankle or foot. In some cases the ulcer has already started to spread and after 4–6 weeks it may be several centimetres in diameter! Two or three adjacent ulcers may join up to form a large sloughing mass that erodes tendons, ligaments and the underlying bone. Even if the bone is not directly involved, X-ray examination may show a marked periosteal reaction to the overlying infection. With time that segment of the bone may become thickened and sclerotic, or there may be erosion of the cortex. With healing, soft-tissue scarring sometimes causes joint contractures at the knee, the ankle or the foot.

Occasionally an invasive squamous cell carcinoma develops in a chronic ulcer.

Treatment

'*Prevention* is better than cure.' For people living or working in the tropics, the chance of infection can be reduced by wearing shoes and any type of covering for the legs. Scratches and abrasions should be cleaned and kept clean until they heal.

Early cases of tropical ulcer may respond to benzylpenicillin or erythromycin given daily for a week. If this is not effective, a broad-spectrum antibiotic will be needed (e.g. a third-generation cephalosporin). Ulcers should be cleansed every day and kept covered with moist or non-adherent dressings. Topical treatment with metronidazole gel is advisable.

Late cases of ulceration will require painstaking cleansing and de-sloughing together with broad-spectrum antibiotics effective against the causative anaerobic Gram-negative organisms as well as secondary infecting microbes cultured from swab samples. Soft-tissue and bone destruction may be severe enough to require extensive debridement and skin-grafting. Occasionally amputation is the best option.

TUBERCULOSIS

Once common throughout the world, tuberculosis showed a steady decline in its prevalence in developed countries during the latter half of the twentieth century, due mainly to the effectiveness of public health programmes, a general improvement in nutritional status and advances in chemotherapy. In the last two decades, however, the annual incidence (particularly of extrapulmonary tuberculosis) has risen again, a phenomenon which has been attributed variously to a general increase in the proportion of elderly people, changes in population movements, the spread of intravenous drug abuse and the emergence of AIDS.

The skeletal manifestations of the disease are seen chiefly in the spine and the large joints, but the infection may appear in any bone or any synovial or bursal sheath. Predisposing conditions include chronic debilitating disorders, diabetes, drug abuse, prolonged corticosteroid medication, AIDS and other disorders resulting in reduced defence mechanisms.

Pathology

Mycobacterium tuberculosis (usually human, sometimes bovine) enters the body via the lung (droplet infection) or the gut (swallowing infected milk products) or, rarely, through the skin. In contrast to pyogenic infection, it causes a granulomatous reaction which is associated with tissue necrosis and caseation.

Primary complex The initial lesion in lung, pharynx or gut is a small one with lymphatic spread to

regional lymph nodes; this combination is the primary complex. Usually the bacilli are fixed in the nodes and no clinical illness results, but occasionally the response is excessive, with enlargement of glands in the neck or abdomen.

Even though there is often no clinical illness, the initial infection has two important sequels: (1) within nodes which are apparently healed or even calcified, bacilli may survive for many years, so that a reservoir exists; (2) the body has been sensitized to the toxin (a positive Mantoux or Heaf test being an index of sensitization) and, should reinfection occur, the response is quite different, the lesion being a destructive one which spreads by contiguity.

Secondary spread If resistance to the original infection is low, widespread dissemination via the bloodstream may occur, giving rise to miliary tuberculosis, meningitis or multiple tuberculous lesions. More often, blood spread occurs months or years later, perhaps during a period of lowered immunity, and bacilli are deposited in extrapulmonary tissues. Some of these foci develop into destructive lesions to which the term 'tertiary' may be applied.

Tertiary lesion Bones or joints are affected in about 5% of patients with tuberculosis (Figure 2.12). There is a predilection for the vertebral bodies and the large synovial joints. Multiple lesions occur in about one-third of patients. In established cases it is difficult to tell whether the infection started in the joint and then spread to the adjacent bone or vice versa; synovial membrane and subchondral bone have a common blood supply and they may, of course, be infected simultaneously.

Once the bacilli have gained a foothold, they elicit a chronic inflammatory reaction. The characteristic microscopic lesion is the tuberculous granuloma (or 'tubercle') – a collection of epithelioid and multinucleated giant cells surrounding an area of necrosis, with round cells (mainly lymphocytes) around the periphery (Figure 2.13).

Within the affected area, small patches of caseous necrosis appear. These may coalesce into a larger yellowish mass, or the centre may break down to form an abscess containing pus and fragments of necrotic bone.

Bone lesions tend to spread quite rapidly. Epiphyseal cartilage is no barrier to invasion and soon the infection reaches the joint. Only in the vertebral bodies, and more rarely in the greater trochanter of the femur or the metatarsals and metacarpals, does the infection persist as a pure chronic osteomyelitis.

If the synovium is involved, it becomes thick and oedematous, giving rise to a marked effusion. A pannus

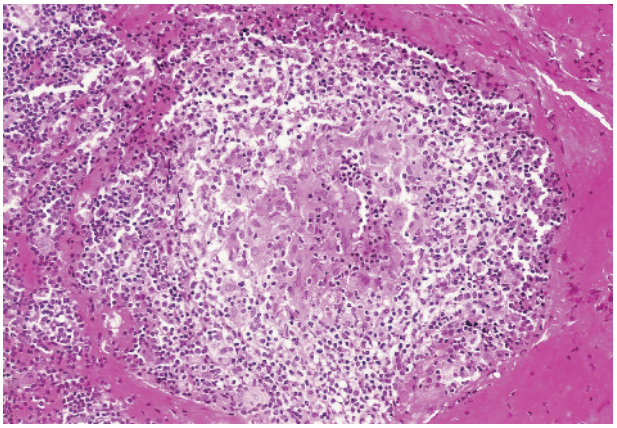
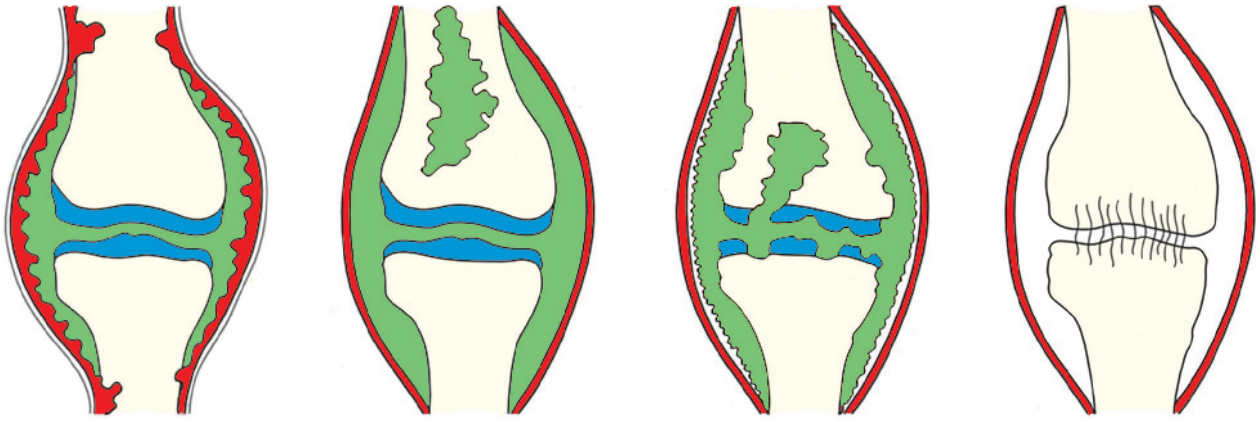


Figure 2.13 Tuberculosis – histology A typical tuberculous granuloma, with central necrosis and scattered giant cells surrounded by lymphocytes and histiocytes.



(a) (b) (c) (d)

Figure 2.12 Tuberculous arthritis – pathology The disease may begin as synovitis (a) or osteomyelitis (b). From either, it can extend to become a true arthritis (c); not all the cartilage is destroyed, and healing is usually by fibrous ankylosis (d).

of granulation tissue may extend from the synovial reflections across the joint; articular cartilage is slowly destroyed, though the rapid and complete destruction elicited by pyogenic organisms does not occur in the absence of secondary infection. At the edges of the joint, along the synovial reflections, there may be active bone erosion. In addition, the increased vascularity causes local osteoporosis.

If unchecked, caseation and infection extend into the surrounding soft tissues to produce a 'cold' abscess ('cold' only in comparison to a pyogenic abscess). This may burst through the skin, forming a sinus or tuberculous ulcer, or it may track along the tissue planes to point at some distant site. Secondary infection by pyogenic organisms is common. If the disease is arrested at an early stage, healing may be by resolution to apparent normality. If articular cartilage has been severely damaged, healing is by fibrosis and incomplete ankylosis, with progressive joint deformity. Within the fibrocaseous mass, mycobacteria may remain imprisoned, retaining the potential to flare up into active disease many years later.

Clinical features

There may be a history of previous infection or recent contact with tuberculosis. The patient, usually a child or young adult, complains of pain and (in a superficial joint) swelling. In advanced cases there may be attacks of fever, night sweats, lassitude and loss of weight. Relatives tell of 'night cries': the joint, splinted by muscle spasm during the waking hours, relaxes with sleep and the inflamed or damaged tissues are stretched or compressed, causing sudden episodes of intense pain. Muscle wasting is characteristic and synovial thickening is often striking (Figure 2.14). Regional lymph nodes may be enlarged and tender. Movements are limited in all directions. As articular erosion progresses the joint becomes stiff and deformed.

In tuberculosis of the spine, pain may be deceptively slight – often no more than an ache when the spine is jarred. Consequently, the patient may not present until there is a visible abscess (usually in the groin or the lumbar region to one side of the midline) or until collapse causes a localized kyphosis. Occasionally, the presenting feature is weakness or instability in the lower limbs.

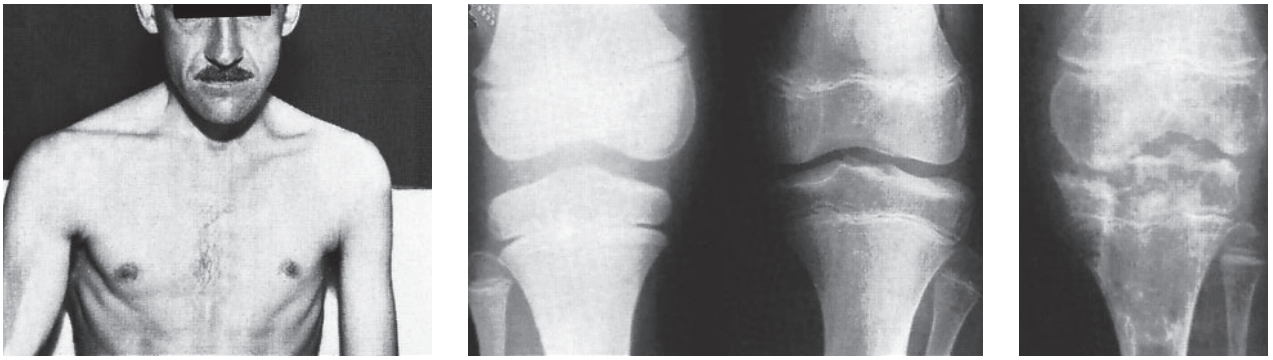
Multiple foci of infection are sometimes found, with bone and joint lesions at different stages of development. This is more likely in people with lowered resistance.

X-ray

Soft-tissue swelling and periarticular osteoporosis are characteristic. The bone ends take on a 'washed-out' appearance and the articular space is narrowed. In children the epiphyses may be enlarged, probably the result of long-continued hyperaemia. Later on there is erosion of the subarticular bone; characteristically this is seen *on both sides of the joint*, indicating an inflammatory process starting in the synovium. Cystic lesions may appear in the adjacent bone ends but there is little or no periosteal reaction. In the spine the characteristic appearance is one of bone erosion and collapse around a diminished intervertebral disc space; the soft-tissue shadows may define a paravertebral abscess.

Investigations

The ESR is usually increased and there may be a relative lymphocytosis. The Mantoux or Heaf test will be positive: these are sensitive but not specific tests; i.e. a negative Mantoux virtually excludes the diagnosis,



(a) (b) (c)

Figure 2.14 Tuberculosis – clinical and X-ray features (a) Generalized wasting used to be a common feature of all forms of tuberculosis. Nowadays, skeletal tuberculosis occurs in deceptively healthy-looking individuals. An early feature is periarticular osteoporosis due to synovitis – the left knee in (b). This often resolves with treatment, but if cartilage and bone are destroyed (c), healing occurs by fibrosis and the joint retains a 'jog' of painful movement.

but a positive test merely indicates tuberculous infection, now or at some time in the past.

If synovial fluid is aspirated, it may be cloudy, the protein concentration is increased and the white cell count is elevated. Acid-fast bacilli are identified in synovial fluid in 10–20% of cases, and cultures are positive in over half. A synovial biopsy is more reliable: sections will show the characteristic histological features and acid-fast bacilli may be identified; cultures are positive in about 80% of patients who have not received antimicrobial treatment.

Diagnosis

Except in areas where tuberculosis is common, diagnosis is often delayed simply because the disease is not suspected. Features that should trigger more active investigation are:

- a long history of pain or swelling
- involvement of only one joint
- marked synovial thickening
- severe muscle wasting
- enlarged and matted regional lymph nodes
- periarticular osteoporosis on X-ray
- a positive Mantoux test.

Synovial biopsy for histological examination and culture is often necessary. Joint tuberculosis must be differentiated from the following.

Transient synovitis This is fairly common in children. At first it seems no different from any other low-grade inflammatory arthritis; however, it always settles down after a few weeks' rest in bed. If the synovitis recurs, further investigation (even a biopsy) may be necessary.

Monarticular rheumatoid arthritis Occasionally, rheumatoid arthritis starts in a single large joint. This is clinically indistinguishable from tuberculosis and the diagnosis may have to await the results of synovial biopsy.

Subacute arthritis Diseases such as amoebic dysentery or brucellosis are sometimes complicated by arthritis. The history, clinical features and pathological investigations usually enable a diagnosis to be made.

Haemorrhagic arthritis The physical signs of blood in a joint may resemble those of tuberculous arthritis. If the bleeding has followed a single recent injury, the history and absence of marked wasting are diagnostic. Following repeated bleeding, as in haemophilia, the clinical resemblance to tuberculosis is closer, but there is also a history of bleeding elsewhere.

Pyogenic arthritis In long-standing cases it may be difficult to exclude an old septic arthritis.

Treatment

REST

Hugh Owen Thomas long ago urged that tuberculosis should be treated by rest – which had to be 'prolonged, uninterrupted, rigid and enforced'. This often involved splintage of the joint and traction to overcome muscle spasm and prevent collapse of the articular surfaces. With modern chemotherapy this is no longer mandatory; rest and splintage are varied according to the needs of the individual patient. Those who are diagnosed and treated early are kept in bed only until pain and systemic symptoms subside, and thereafter are allowed restricted activity until the joint changes resolve (usually 6 months to a year). Those with progressive joint destruction may need a longer period of rest and splintage to prevent ankylosis in a bad position; however, as soon as symptoms permit, movements are again encouraged.

CHEMOTHERAPY

The most effective treatment is a combination of antituberculous drugs, which should always include rifampicin and isoniazid. During the last decade the incidence of drug resistance has increased and this has led to the addition of various 'potentiating' drugs to the list. The following is one of several recommended regimens.

Initial, '*intensive phase treatment*', consists of isoniazid 300–400 mg, rifampicin 450–600 mg and fluoroquinolones 400–600 mg daily for 5–6 months. All replicating sensitive bacteria are likely to be killed by this bactericidal attack. This is followed by a '*continuation phase treatment*' lasting 9 months, the purpose of which is to eliminate the 'persisters', slow-growing, intermittently-growing, dormant or intracellular mycobacteria. This involves the use of isoniazid and pyrazinamide 1500 mg per day for 4½ months and isoniazid and rifampicin for another 4½ months. Then a '*prophylactic phase*', consisting of isoniazid and ethambutol 1200 mg per day for a further 3–4 months.

During the entire treatment period, drugs and dosage may have to be adjusted and modified, depending on the individual patient's age, size, general health and drug reactions.

OPERATION

Operative drainage or clearance of a tuberculous focus is seldom necessary nowadays. However, a cold abscess may need immediate aspiration or draining.

Once the condition is controlled and arthritis has completely subsided, normal activity can be resumed, though the patient must report any renewed symptoms. If, however, the joint is painful and the articular surface is destroyed, arthrodesis or replacement arthroplasty may be considered. The longer the period

of quiescence, the less the risk of reactivation of the disease; there is always some risk and it is essential to give chemotherapy for 3 months before and after the operation.

BRUCELLOSIS

Brucellosis is an unusual but nonetheless important cause of subacute or chronic granulomatous infection in bones and joints. Three species of organism are seen in humans: *Brucella melitensis*, *Brucella abortus* (from cattle) and *Brucella suis* (from pigs). Infection usually occurs from drinking unpasteurized milk or from coming into contact with infected meat (e.g. among farmers and meat packers). In the past it has been more common in countries around the Mediterranean and in certain parts of Africa and India. About 50% of patients with chronic brucellosis develop arthritis.

Pathology

The organism enters the body with infected milk products or, occasionally, directly through the skin or mucosal surfaces. It is taken up by the lymphatics and then carried by the blood stream to distant sites. Foci of infection may occur in bones (usually the vertebral bodies) or in the synovium of the larger joints. The characteristic lesion is a chronic inflammatory granuloma with round-cell infiltration and giant cells. There may be central necrosis and caseation leading to abscess formation and invasion of the surrounding tissues.

Clinical features

The patient usually presents with fever, headache and generalized weakness, followed by joint pains and backache. The initial illness may be acute and alarming; more often it begins insidiously and progresses until the symptoms localize in a single large joint (usually the hip or knee) or in the spine. The joint becomes painful, swollen and tender; movements are restricted in all directions. If the spine is affected, there is usually local tenderness and back movements are restricted.

The systemic illness follows a fluctuating course, with alternating periods of fever and apparent improvement (hence the older term 'undulant fever'). Diagnosis is often long delayed and may not be resolved until destructive changes are advanced.

X-rays

The picture is that of a subacute arthritis, with loss of articular space, slowly progressive bone erosion and periarticular osteoporosis. In the spine there may be destruction and collapse of adjacent vertebral bodies with obliteration of the disc.

Investigations

A positive agglutination test (titre above 1/80) is diagnostic. Joint aspiration or biopsy may allow the organism to be cultured and identified.

Diagnosis

Diagnosis is usually delayed while other types of subacute arthritis are excluded.

Tuberculosis and brucellosis have similar clinical and radiological features. The distinction is often difficult and may have to await the results of agglutination tests, synovial biopsy and bacteriological investigation.

Reiter's disease and other forms of reactive arthritis often follow an initial systemic illness. However, fever is not so marked and joint erosion is usually late and mild.

Treatment

ANTIBIOTICS

The infection usually responds to a combined onslaught with tetracycline and streptomycin for 3–4 weeks. Alternative drugs, which are equally effective and which may be used as 'combination therapy', are rifampicin and the newer cephalosporins.

Operation An abscess will need drainage, and necrotic bone and cartilage should be meticulously excised. If the joint is destroyed, arthrodesis or arthroplasty may be necessary once the infection is completely controlled.

LEPROSY

Leprosy is a mildly infectious chronic inflammatory disease caused by acid-fast *Mycobacterium leprae*. It is characterized by granulomatous lesions in the peripheral nerves, the skin and the mucosa of the upper respiratory tract.

Leprosy was once common throughout the world. Today it is rarely seen outside parts of South Asia, Africa, Latin America and some of the Pacific Islands. While the disease is easily cured with drugs, its crippling effects persist in a cumulative number of people.

The infection is acquired mainly by respiratory transmission; unbroken skin to skin contact is thought not to be dangerous. Several years may elapse before clinical features appear.

Pathology

Most people infected with *Mycobacterium leprae* develop protective immunity and get rid of the infection.

Some develop a few skin lesions, appearing as vague hypopigmented macules (*indeterminate leprosy*), that recover spontaneously. If the condition progresses, it takes one of several forms, depending on the host's immune response.

Tuberculoid leprosy occurs where there is delayed type hypersensitivity (DTH) to *Mycobacterium leprae* antigens, combined with some decrease in cell-mediated immunity (CMI). The granuloma in tuberculoid leprosy is focal and circumscribed and is made up of epithelioid cells, with a few scattered giant cells and a cuff of lymphocytes, very similar to tuberculosis.

Lepromatous leprosy is seen in patients who are unable to mount effective CMI against *Mycobacterium leprae*. Here the granuloma is diffuse and extensive and it consists of macrophages, many loaded with acid-fast bacilli. There may be a sprinkling of round cells in the lepromatous granuloma. The entire body skin may thus be affected.

Borderline types are intermediate forms that show some features of both of the above conditions. Without treatment, they tend to progress increasingly towards the lepromatous form.

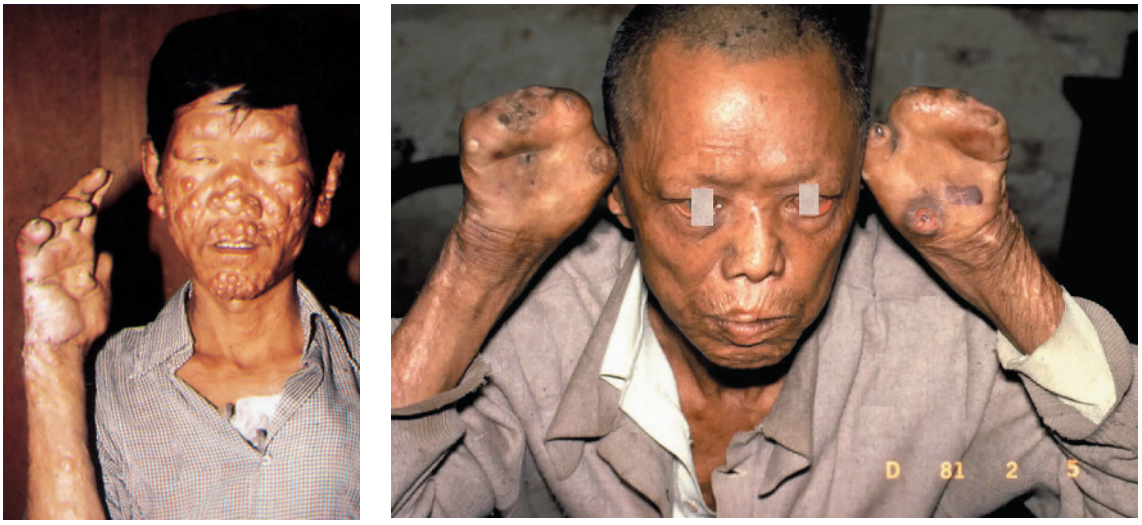
Peripheral nerves are always affected in leprosy. Dermal nerve twigs, cutaneous nerves as well as major nerve trunks may thus be involved. The affected nerves become thickened. Besides the granuloma there is hypertrophy of the epineurium and perineurium, demyelination, axonal degeneration and endoneurial fibrosis. A thickened nerve trunk may be strangulated by its own sheath or by the rigid walls of a fibro-osseous tunnel through which it passes (e.g. the ulnar nerve at the elbow). Sometimes, a tuberculoid granuloma in a nerve undergoes caseation. An important factor contributing to nerve damage is that medication is less likely to reach the segment of the nerve thus rendered ischaemic.

The chronic course of leprosy is often punctuated by acute inflammatory episodes – so-called '*reactions*' – which are due to the deposition of immune complexes (erythema nodosum leprosum or ENL or Type II reaction) or due to an increase in CMI and DTH levels (reversal reaction or RR or Type I reaction). Reactions occurring in the nerves (acute neuritis) greatly increase the risk of nerve damage.

Clinical features

Hypopigmented skin patches with impaired sensibility develop in all types of leprosy. Thickened cutaneous nerves may be seen and thickened nerve trunks may be felt where they are superficial, especially where they cross a bone (typically behind the medial condyle of the humerus at the elbow). Irrecoverable nerve damage with characteristic patterns of muscle weakness and deformities of the hands and feet may also be seen (Figure 2.15). Trophic ulcers, causing progressive destruction of the affected part, appear in the hands and feet.

Skin lesions in *tuberculoid leprosy* are sparse, well-demarcated, hypopigmented and anaesthetic. In contrast, in *lepromatous leprosy*, the skin is affected diffusely and extensively and the lesions present as multiple, symmetrically distributed macular patches with some sensory impairment. Plaques and nodules develop in advanced stages. Coarsening of the facial skin and loss of eyebrows may produce typical leonine features. Lepromatous ulceration of the nasal mucosa leads to destruction of the nasal septum and nasal deformity.



(a) (b)

Figure 2.15 Leprosy – late features (a) Patient showing typical ulnar claw-hand deformity. (b) This patient was even worse off, having lost all the fingers of both hands.

Peripheral nerves are affected extensively in *lepromatous leprosy* whereas in *tuberculoid leprosy* the neural lesions are few and focal in distribution. Cutaneous nerves as well as major nerve trunks of the upper and lower limbs are usually involved. Except for the Vth and VIIth nerves, the cranial nerves are not affected. Clinical defects in nerve function appear early in *tuberculoid leprosy* but much later in *lepromatous leprosy.*

Nerve lesions in tuberculoid leprosy may undergo caseation and liquefaction resulting in an intraneural 'cold abscess' mimicking an intraneural tumour, or the pus may break through the epineurium to present as a chronic collar-stud abscess.

Diagnosis

In countries where the disease is common the clinical diagnosis is seldom in doubt. Suggestive signs are the appearance of skin lesions with loss of sensibility, palpably or even visibly thickened nerves which may also be tender, areas of anaesthesia, chronic ulcers of the feet and typical deformities of hands and feet due to muscle weakness and imbalance. In countries where the disease is not endemic, diagnosis may have to await the results of skin smear examination, serological tests and skin or nerve biopsy.

Patterns of nerve involvement

Nerve trunks of the upper limbs are involved more often than those of the lower limbs. There is a pattern in the selection, site of involvement, risk of damage and chances of recovery (see Table 2.2). In the upper limb ulnar nerve paralysis is the most common and combined ulnar and median nerve paralysis is seen less frequently. Occasionally, triple nerve paralysis (paralysis of ulnar, median and radial nerves) may occur. Any other pattern is extremely rare.

Treatment

For purposes of treatment, patients are categorized as having *paucibacillary* (cases of indeterminate and tuberculoid leprosy) or *multibacillary* (cases of lepromatous and borderline leprosy) leprosy.

MULTIDRUG THERAPY

Combined chemotherapy with rifampicin as one of the drugs is the mainstay of treatment; however, the choice of drugs and duration of treatment depend on the type of disease. Following the recommendations of the World Health Organization, patients with *paucibacillary disease* are treated with rifampicin 600 mg once monthly and dapsone 100 mg once daily, for 6 months; patients with *multibacillary disease* are given rifampicin 600 mg and clofazi mine 300 mg once monthly and dapsone 100 mg and clofazimine 50 mg once daily, for 12 months. Reactions, especially acute neuritis, are treated with anti-inflammatory medication, of which prednisolone is the most important, and other supportive therapy.

NERVE DECOMPRESSION

Surgical decompression of a nerve trunk is sometimes required in order to improve perfusion of the nerve and allow the anti-leprosy and anti-inflammatory drugs to reach the affected segment and thus prevent or abort nerve damage. Surgical decompression is indicated: (a) in acute neuritis when, even while under treatment with corticosteroids, there is increasing neurological deficit; and (b) in cases of severe, unresponsive nerve pain, for relief of pain. Decompression involves tunnel release (often with excision of the medial epicondyle for the ulnar nerve) combined with incision of the epineurium over the entire sclerosed segment of the nerve. *Stripping the epineurium should not be done.*

TREATMENT OF NERVE ABSCESS

Cold abscesses associated with deteriorating neurological function and those that are likely to burst through the skin need to be excised or surgically evacuated. If there is no associated neural deficit, it is not necessary to intervene immediately, provided the patient can be reviewed periodically.

Table 2.2 Features of nerve trunk involvement in leprosy

Nerve affected	Preferred site	Involvement[a]	Motor paralysis	Recovery
Ulnar[c]	Above elbow/wrist	++++	++++	+
Median	Above wrist	++	++	++
Common peroneal	Back of knee	+++	+	++
Tibia	Behind ankle	+++	+++	[b]
Radial	Cutaneous division	+++	NA	NA
	Radial groove	++	(Forearm muscles only)	+++

[a] Thickening; [b] tenderness/pain; [c] most commonly involved nerve trunk; + uncommon; ++ common; +++ quite common; ++++ very common; NA, not applicable.

MANAGEMENT OF RESIDUAL PARALYSIS AND TROPHIC LESIONS

The long-term neuropathic complications of leprosy are dealt with in Chapter 11. The notorious deformities and disablement result from: (a) *local leprous granulomas* (as in the face); (b) *damage to nerves* of the hands and feet and consequent muscle paralysis; and (c) so-called '*trophic lesions*' (ulcers, shortening of digits and mutilations) arising from injuries to insensitive hands and feet. These conditions are prevented by early treatment of the disease, adequate treatment of neuritis and protection of anaesthetic hands and feet.

Paresis and established deformities can usually be corrected or at least improved by surgery. Although this is done mainly to improve function, restoration of normal appearance is also important for leprosy patients. Deformities such as claw fingers and drop foot stigmatize affected individuals as 'leprosy patients', with dire social consequences.

Individuals requiring surgery should have had antileprosy treatment and should not have had acute neuritis of any nerve trunk for at least 6 months prior to surgery. They must be well motivated and there should be proper pre-operative preparation with appropriate physiotherapy. Absence of facilities for pre- and postoperative therapy is an absolute contraindication for corrective surgery.

MYCOTIC INFECTIONS

Mycotic or fungal infection causes an indolent granulomatous reaction, often leading to abscess formation, tissue destruction and ulceration. When the musculoskeletal system is involved, it is usually by direct spread from the adjacent soft tissues. Occasionally, however, a bone or joint may be infected by haematogenous spread from a distant site.

These disorders are conveniently divided into 'superficial' and 'deep' infections.

Superficial mycoses These are primarily infections of the skin or mucous surfaces which spread into the adjacent soft tissues and bone. The more common examples are the *maduromycoses* (a group consisting of several species), *Sporothrix* and various species of *Candida*.

The *actinomycoses* are usually included with the superficial fungal infections. The causal organisms, of which *Actinomyces israelii* is the commonest in humans, are not really fungi but anaerobic bacilli with fungus-like appearance and behaviour.

Deep mycoses This group comprises infections by *Blastomyces*, *Histoplasma*, *Coccidioides*, *Cryptococcus*, *Aspergillus* and other rare fungi. The organisms, which occur in rotting vegetation and bird droppings, gain entry through the lungs and, in humans, may cause an influenza-like illness. Bone or joint infection is uncommon except in patients with compromised host defences.

MADUROMYCOSIS

This chronic fungal infection is seen mainly in northern Africa and the Indian subcontinent. The organisms usually enter through a cut in the foot; from there they spread through the subcutaneous tissues and along the tendon sheaths. The bones and joints are infected by direct invasion; local abscesses form and break through the skin as multiple sinuses. The patient may present at an early stage with a tender subcutaneous nodule (when the diagnosis is seldom entertained); more often he or she is seen when the foot is swollen and indurated, with discharging sinuses and ulcers (Figure 2.16). *X-rays* may show multiple bone cavities or progressive bone destruction. The organism can be identified in the sinus discharge or in tissue biopsies.

Treatment is unsatisfactory as there is no really effective chemotherapy. Intravenous amphotericin B is advocated, but it is fairly toxic and causes side effects such as headaches, vomiting and fever. Necrotic tissue should be widely excised. Even then it is sometimes difficult to stop further invasion, and amputation is sometimes necessary.

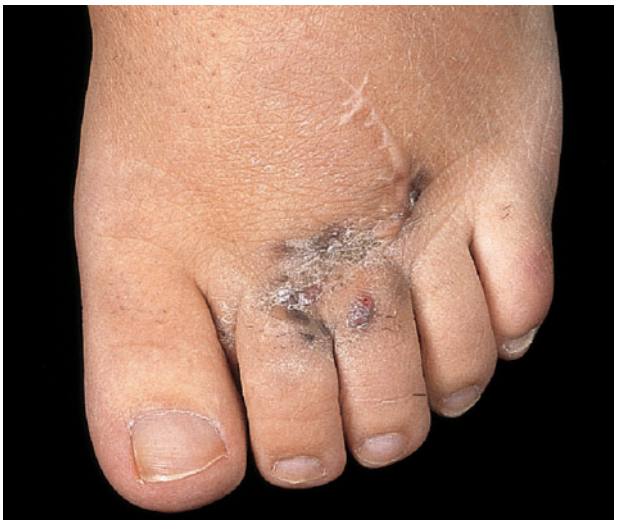


Figure 2.16 Maduromycosis This Mediterranean market-worker was perpetually troubled by tiny abscesses and weeping sinuses in her foot. X-rays showed that bone destruction had already spread to the tarsal bones, and after 2 years of futile treatment the foot had to be amputated.

Candidiasis

Candida albicans is a normal commensal in humans and it often causes superficial infection of the skin or mucous membranes. Deep and systemic infections are rare except under conditions of immunosuppression.

Candida osteomyelitis and arthritis may follow direct contamination during surgery or other invasive procedures such as joint aspiration or arthroscopy. The diagnosis is usually made only after tissue sampling and culture.

Treatment consists of thorough joint irrigation and curettage of discrete bone lesions, together with intravenous amphotericin B.

Actinomycosis

Infection is usually by *Actinomyces israelii*, an anaerobic Gram-positive bacillus. Although rare, it is important that it should be diagnosed because the organism is sensitive to antibiotics.

The most common site of infection is the mandible (from the mouth and pharynx), but bone lesions are also seen in the vertebrae (spreading from the lung or gut) and the pelvis (spreading from the caecum or colon). Peripheral lesions may occur by direct infection of the soft tissues and later extension to the bones. There may be a firm, tender swelling in the soft tissues, going on to form an abscess and one or more chronic discharging sinuses. *X-rays* may show cyst-like areas of bone destruction. The organism can be readily identified in the sinus discharge, but only on anaerobic culture.

Treatment, by large doses of benzylpenicillin G, tetracycline or erythromycin, has to be continued for several months.

The deep mycoses

Histoplasmosis, blastomycosis and coccidioidomycosis are rare causes of bone and joint infection, but they should always be considered in patients on immunosuppressive therapy who develop arthritis of one of the large joints or osteomyelitis in an unusual site. Diagnosis is usually delayed and often involves specialized microbiological investigations to identify the organism.

Treatment with intravenous amphotericin B is moderately effective. Operation may be necessary to drain an abscess or to remove necrotic tissue.

HYDATID DISEASE

Hydatid disease is caused by the tapeworm *Echinococcus*. Parasitic infestation is common among sheep farmers, but bone lesions are rare.

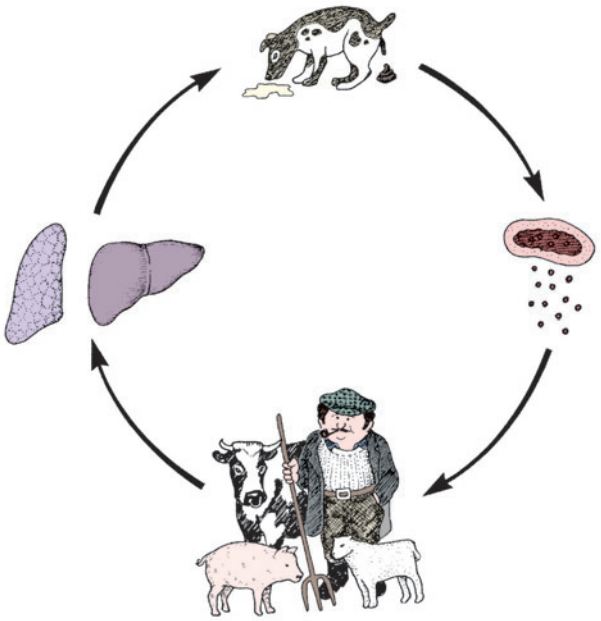


Figure 2.17 Hydatid disease The life cycle of the tapeworm which causes hydatid disease.

The organism, a cestode worm, has a complicated life cycle (Figure 2.17). The definitive host is the dog or some other carnivore that carries the tapeworm in its bowel. Segments of worm and ova pass out in the faeces and are later ingested by one of the intermediate hosts – usually sheep or cattle or man. Here the larvae are carried via the portal circulation to the liver, and occasionally beyond to other organs, where they produce cysts containing numerous scolices. Infested meat is then eaten by dogs (or humans), giving rise to a new generation of tapeworm.

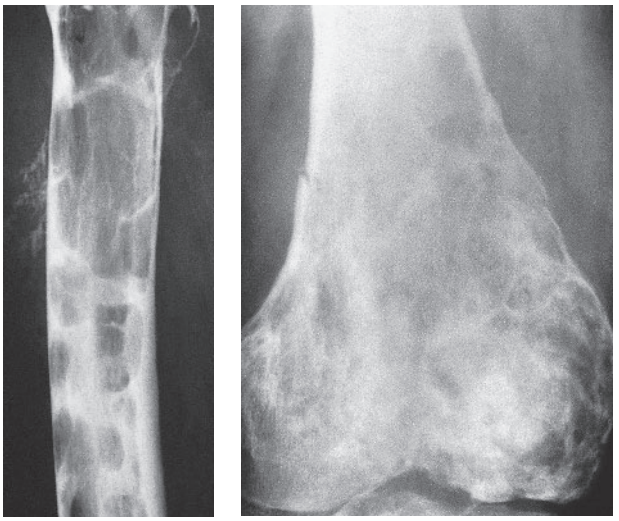
Scolices carried in the bloodstream occasionally settle in bone and produce hydatid cysts that slowly enlarge with little respect for cortical or epiphyseal boundaries. The bones most commonly affected are the vertebrae, pelvis, femur, scapula and ribs.

Clinical features

The patient may complain of pain and swelling, or may present for the first time with a pathological fracture or compression of the spinal cord. Infestation sometimes starts in childhood but the cysts take so long to enlarge that clinical symptoms and signs may not become apparent for many years. The diagnosis is more likely if the patient comes from a sheep-farming district.

Imaging

X-rays show solitary or multiloculated bone cysts, but only moderate expansion of the cortices (Figure 2.18). However, cortical thinning may lead to a pathological fracture. In the spine, hydatid disease may involve



(a) (b)

Figure 2.18 Hydatid disease of bone Two examples of hydatid involvement of bone: there is no expansion of the cortex in (a) and very little in (b).

adjacent vertebrae, with large cysts extending into the paravertebral soft tissues. These features are best seen on *CT* and *MRI*, which should always be performed if operative excision of the lesion is contemplated.

Investigations

Casoni's (complement fixation) test may be positive, especially in long-standing cases.

Diagnosis

Hydatid disease must be included in the differential diagnosis of benign and malignant bone cysts and cyst-like tumours. If the clinical and radiological features are not conclusive, needle biopsy should be considered, though there is a risk of spreading the disease.

Treatment

The antihelminthic drug albendazole is moderately effective in destroying the parasite. It has to be given in repeated courses: a recommended programme is oral administration of 10 mg per kg per day for 3 weeks, repeated at least 4 times with a 1-week 'rest' between courses. Liver, renal and bone marrow function should be monitored during treatment.

However, the bone cysts do not heal and recurrence is common. The indications for surgery are continuing enlargement or spread of the lesion, a risk of fracture, invasion of soft tissues and pressure on important structures. Curettage and bone grafting will lessen the risk of pathological fracture; at operation the cavity can be 'sterilized' with copious amounts of hypertonic saline, alcohol or formalin to lessen the risk of recurrence.

Radical resection, with the margin at least 2 cm beyond the cyst, is more certain, but also much more challenging. In a long bone, the space can sometimes be filled with a tumour-prosthesis, to include an arthroplasty if necessary. Large cysts of the vertebral column, or the pelvis and hip joint, are particularly difficult to manage in this way and in some cases surgical excision is simply impractical or impossible.

FINAL COMMENT

Infections are severe clinical entities that need to be considered in many clinical scenarios. When affecting bones or joints, and especially when implants are involved, microorganisms may attach and proliferate until the point of severely damaging the tissue and the general health of the patient and eventually proving fatal. Even if cured, an infection may seriously affect the appropriate function of the bone and joint, and cause long-term or even permanent disability.

Early clinical suspicion, adequate aetiological diagnosis, and properly staged treatment that usually includes surgery, antibiotics, and other actions, are crucial to control and eventually heal these complex diseases.

Future management of increasingly complex infections will require a deep knowledge of available diagnostic and therapeutic options and developments. From basic clinical reasoning to sophisticated laboratory tools, from appropriate surgical decisions to specific antibiotic regimes, a multidisciplinary approach is a major asset to successfully orient musculoskeletal infections.

FURTHER READING

Cierny G 3rd, Mader JT, Penninck JJ. A clinical staging system for adult osteomyelitis. *Clin Orthop Relat Res* 2003; **414**: 7–24.

Dartnell J, Ramachandran M, Katchburian M. Haematogenous acute and subacute paediatric osteomyelitis: a systematic review of the literature. *J Bone Joint Surg Br.* 2012; **94**(5): 584–95.

Donlan RM, Costerton JW. Biofilms: survival mechanisms of clinically relevant microorganisms. *Clin Microbiol Rev* 2002; **15**: 167–93.

Gristina AG. Biomaterial-centered infection: microbial adhesion versus tissue integration. *Science* 1987; **237**(4822): 1588–95.

Perez-Jorge C, Gómez-Barrena E, Horcajada JP, *et al.* Drug treatments for prosthetic joint infections in the era of multidrug resistance. *Expert Opin Pharmacother* 2016 Apr 7. [Epub ahead of print] PMID: 27054293. Available at: http://dx.doi.org/10.1080/14656566.2016.1176142 [Accessed 7 February 2017].

Tande AJ, Patel R. Prosthetic joint infection. *Clin Microbiol Rev* 2014; **27**(2): 302–45.

Thang SP, Tong AK, Lam WW, *et al.* SPECT/CT in musculoskeletal infections. *Semin Musculoskelet Radiol* 2014; **18**(2): 194–202.

Tsukayama DT, Estrada R, Gustilo RB. Infection after total hip arthroplasty: a study of the treatment of one hundred and six infections. *J Bone Joint Surg* 1996; **78A**: 512–23.

Taylor & Francis
Taylor & Francis Group
http://taylorandfrancis.com

Inflammatory rheumatic disorders

3

Christopher Edwards

The term 'inflammatory rheumatic disorders' covers a number of diseases that cause chronic pain, stiffness and swelling around joints and tendons. In addition, they are commonly associated with extra-articular features including skin rashes and inflammatory eye disease. Individuals with these diseases tend to die younger than their peers as a result of the effects of chronic inflammation. Many – perhaps all – are due to a faulty immune or inflammatory reaction resulting from a combination of environmental exposures against a background of genetic predisposition.

RHEUMATOID ARTHRITIS

Rheumatoid arthritis (RA) is the most common cause of chronic inflammatory joint disease. The most typical features are a symmetrical polyarthritis and tenosynovitis, morning stiffness, elevation of the erythrocyte sedimentation rate (ESR) and the appearance of autoantibodies (rheumatoid factor (RF) and anti-citrullinated peptide antibodies (ACPAs)) in the serum. Rheumatoid arthritis is a systemic disease and changes can be widespread in a number of tissues. Individuals with RA tend to die younger than their peers as a result of the effects of chronic inflammation on a number of organ systems. Chief among these is early ischaemic heart disease secondary to the effects of inflammation on the cardiovascular system.

The reported prevalence of RA in most populations is 1–2%, with a peak incidence in the fourth or fifth decades. Women are affected three times more commonly than men. Both the prevalence and the clinical expression vary between populations; the disease is more common (and generally more severe) in Caucasians living in the urban communities of Europe and North America than in the rural populations of Africa.

Cause

The cause of RA is still incompletely worked out. However, a great deal is now known about the circumstances in which RA develops, and hypotheses about its aetiology and pathogenesis have been suggested. Important factors in the evolution of RA are: (1) genetic susceptibility; (2) an immunological reaction, possibly involving a foreign antigen, preferentially focused on synovial tissue; (3) an inflammatory reaction in joints and tendon sheaths; (4) the appearance of rheumatoid factors (RF) and anti-citrullinated antibodies (anti-CCP or ACPA) in the blood and synovium; (5) perpetuation of the inflammatory process; and (6) articular cartilage destruction.

Genetic susceptibility A genetic association is suggested by the fact that RA is more common in first-degree relatives of patients than in the population at large; furthermore, twin studies have revealed a concordance rate of around 30% if one of the pair is affected. The human leucocyte antigen (HLA) DR4 occurs in about 70% of people with RA, compared to a frequency of less than 30% in normal controls. HLA-DR4 is encoded in the major histocompatibility complex (MHC) region on chromosome 6. There are strong associations between HLA-DR4 and RA. In particular, a key structural conformation within the HLA-DR4 binding groove called the 'shared epitope' seems important. This may suggest that a particular antigen that fits into this may be playing a part.

HLA Class II molecules appear as surface antigens on cells of the immune system (B lymphocytes, macrophages, dendritic cells), which can act as antigen-presenting cells (APCs). In some T-cell immune reactions, the process is initiated only when the antigenic peptide is presented in association with a specific HLA allele. It has been suggested that this is the case in people who develop RA; the idea is even more attractive if one proposes that the putative antigen has a special affinity for synovial tissue. So far no such antigen has been discovered.

The inflammatory reaction Once the APC/T-cell interaction is initiated, various local factors come into play and lead to a progressive enhancement of the immune response. There is a marked proliferation of

cells in the synovium, with the appearance of new blood vessel formation. Immune cells coordinate their action by the use of 'short-range hormones' (cytokines), which can activate inflammatory cells such as macrophages and B cells. Some cytokines, called chemokines, attract other inflammatory cells to the area.

Over recent years it has become clear that certain cytokines are important in RA. These include tumour necrosis factor (TNF), interleukin-1 (IL-1) and interleukin-6 (IL-6). The resulting synovitis, both in joints and in tendon sheath linings, is the hallmark of early RA.

Rheumatoid factor B-cell activation in RA leads to the production of anti-IgG autoantibodies, which are detected in the blood as 'rheumatoid factor' (RF). Low levels of RF can be found in many 'normal' individuals but, when the levels are high, an inflammatory disease is likely. Other autoimmune conditions such as systemic lupus erythematosus (SLE) and Sjögren's syndrome are also associated with the presence of RF.

In recent years other autoantibodies associated with RA have been identified. The most important are anti-cyclic citrullinated peptide antibodies (anti-CCP). The presence of anti-CCP is very specific for RA. Patients with a positive RF test tend to be more severely affected than those with a negative test.

Chronic synovitis and joint destruction Chronic rheumatoid synovitis is associated with the production of proteolytic enzymes, prostaglandins and the cytokines TNF and IL-1. Immune complexes are deposited in synovial joints, where they appear to augment the inflammatory process. This combination of factors leads to depletion of the cartilage matrix and, eventually, damage to cartilage and underlying bone. Vascular proliferation and osteoclastic activity, most marked at the edges of the articular surface, may contribute further to cartilage destruction and periarticular bone erosion.

Pathology

Rheumatoid arthritis is a systemic disease but the most characteristic lesions are seen in the synovium or within rheumatoid nodules. The synovium is engorged with new blood vessels and packed full of inflammatory cells.

JOINTS AND TENDONS

The pathological changes, if unchecked, proceed in four stages (Figure 3.1). Previously it was felt that having gone through these stages the disease activity could be 'burnt out'. This does not appear to be the case. In any one joint, features of different stages can be occurring simultaneously and even when joints are very badly destroyed the ongoing inflammation can continue to seriously damage systemic health by accelerating other disease processes such as ischaemic heart disease.

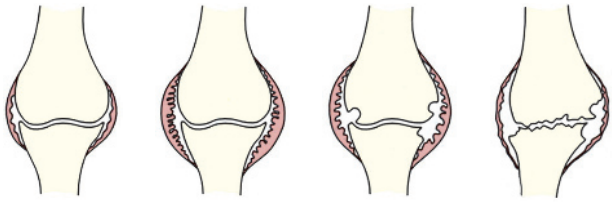


Figure 3.1 Rheumatoid arthritis – pathology
(a) Stage 1 – pre-clinical. (b) Stage 2 – synovitis and joint swelling. (c) Stage 3 – early joint destruction with peri-articular erosions. (d) Stage 4 – advanced joint destruction and deformity.

Stage 1: Pre-clinical Well before RA becomes clinically apparent, the immune pathology is already beginning. Raised ESR, C-reactive protein (CRP) and RF may be detectable years before the first diagnosis.

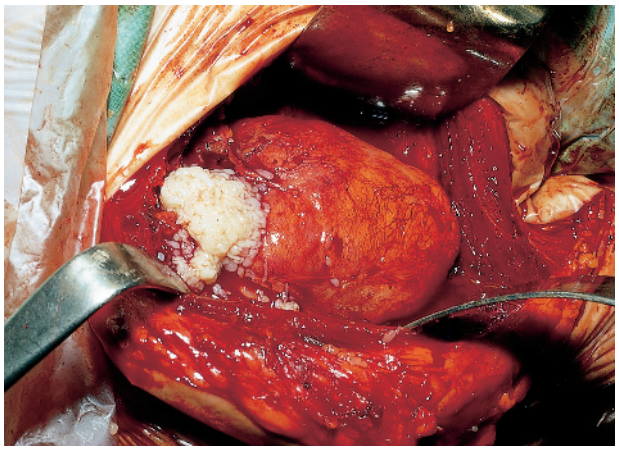
Stage 2: Synovitis Early changes are vascular congestion with new blood vessel formation, proliferation of synoviocytes and infiltration of the subsynovial layers by polymorphs, lymphocytes and plasma cells (Figure 3.2). There is thickening of the capsular structures, villous formation of the synovium and a cell-rich effusion into the joints and tendon sheaths. Although painful, swollen and tender, these structures are still intact and mobile, and the disorder is potentially reversible.

Stage 3: Destruction Persistent inflammation causes joint and tendon destruction. Articular cartilage is eroded, partly by proteolytic enzymes, partly by vascular tissue in the folds of the synovium, and partly due to direct invasion of the cartilage by a pannus of granulation tissue creeping over the articular surface. At the margins of the joint, bone is eroded by tissue invasion and osteoclastic resorption. Similar changes occur in tendon sheaths, causing tenosynovitis, invasion of the collagen bundles and, eventually, partial or complete rupture of tendons. A synovial effusion, often containing copious amounts of fibrinoid material, produces swelling of the joints, tendons and bursae.

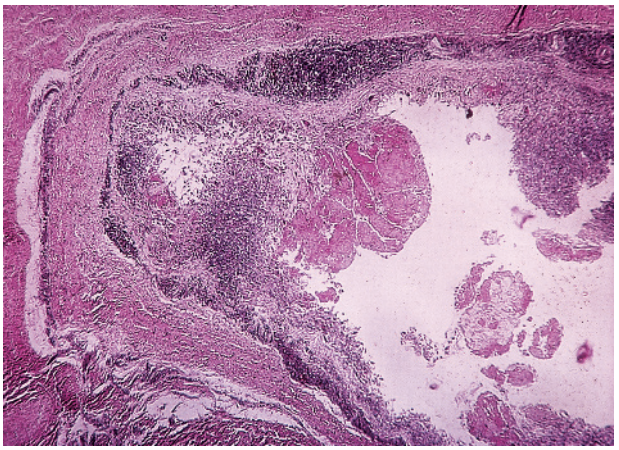
Stage 4: Deformity The combination of articular destruction, capsular stretching and tendon rupture leads to progressive instability and deformity of the joints. The inflammatory process usually continues but the mechanical and functional effects of joint and tendon disruption now become vital.

EXTRA-ARTICULAR TISSUES

Rheumatoid nodules The rheumatoid nodule is a small granulomatous lesion consisting of a central necrotic zone surrounded by a radially disposed



(a)



(b)

Figure 3.2 Rheumatoid synovitis (a) The macroscopic appearance of rheumatoid synovitis with fibrinoid material oozing through a rent in the capsule. (b) Histology shows proliferating synovium with round-cell infiltration and fibrinoid particles in the joint cavity (×120).

palisade of local histiocytes, and beyond that by inflammatory granulation tissue. Nodules occur under the skin (especially over bony prominences), in the synovium, on tendons, in the sclera and in many of the viscera.

Lymphadenopathy Not only the nodes draining inflamed joints, but also those at a distance such as the mediastinal nodes, can be affected. This, as well as a mild *splenomegaly*, is due to hyperactivity of the reticuloendothelial system. More severe splenomegaly can also be associated with neutropaenia as part of *Felty's syndrome*.

Vasculitis This can be a serious and life-threatening complication of RA. Involvement of the skin, including nailfold infarcts, is common but organ infarction can occur.

Muscle weakness Muscle weakness is common. It may be due to a generalized *myopathy* or *neuropathy*, but it is important to exclude spinal cord disease or cord compression due to vertebral displacement (atlantoaxial subluxation). Sensory changes may be part of a neuropathy, but localized sensory and motor symptoms can also result from *nerve compression* by thickened synovium (e.g. carpal tunnel syndrome).

Visceral disease The lungs, heart, kidneys, gastrointestinal tract and brain are sometimes affected. *Ischaemic heart disease* and *osteoporosis* are common complications.

Clinical features

The onset of RA is usually insidious, with symptoms emerging over a period of months. Occasionally the disease starts quite suddenly.

In the early stages the picture is mainly that of a polysynovitis, with soft-tissue swelling and stiffness (Figure 3.3). Typically, a woman of 30–40 years complains of pain, swelling and loss of mobility in the proximal joints of the fingers. There may be a previous history of 'muscle pain', tiredness, loss of weight and a general lack of well-being. As time passes, the symptoms 'spread' to other joints – the wrists, feet, knees and shoulders in order of frequency. Another classic feature is generalized stiffness after periods of inactivity, and especially after rising from bed in the early morning. This early morning stiffness typically lasts longer than 30 minutes.

Physical signs may be minimal, but usually there is symmetrically distributed swelling and tenderness of the metacarpophalangeal joints, the proximal interphalangeal joints and the wrists. Tenosynovitis is common in the extensor compartments of the wrist and the flexor sheaths of the fingers; it is diagnosed by feeling thickening, tenderness and crepitation over the back of the wrist or the palm while passively moving the fingers. If the larger joints are involved, local warmth, synovial hypertrophy and intra-articular effusion may be more obvious. Movements are often limited but the joints are still stable and deformity is unusual.

In the later stages joint deformity becomes increasingly apparent and the acute pain of synovitis is replaced by the more constant ache of progressive joint destruction. The combination of joint instability and tendon rupture produces the typical 'rheumatoid' deformities: ulnar deviation of the fingers, radial and volar displacement of the wrists, valgus knees, valgus feet and clawed toes. Joint movements are restricted and often very painful. About a third of all patients develop pain and stiffness in the cervical spine.

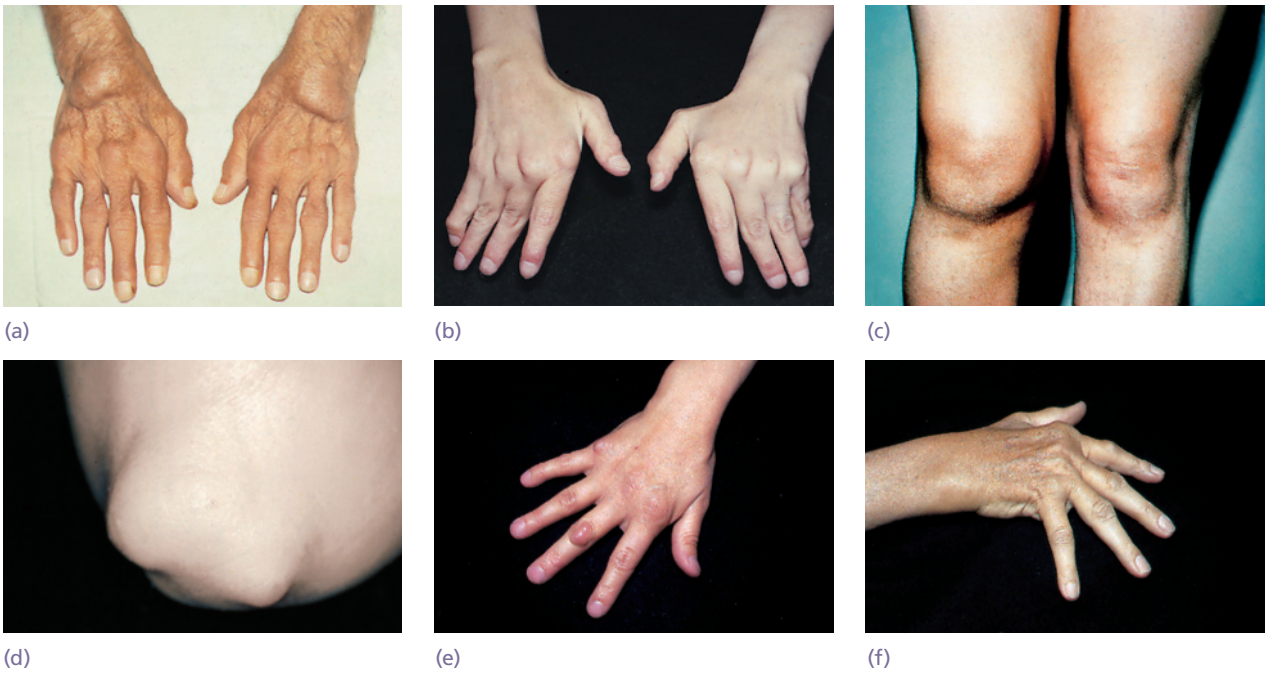


Figure 3.3 Rheumatoid arthritis – clinical features (a) Early features of swelling and stiffness of the proximal finger joints and the wrists. (b) The late hand deformities are so characteristic as to be almost pathognomonic. (c) Occasionally rheumatoid disease starts with synovitis of a single large joint (in this case the right knee). Extra-articular features include subcutaneous nodules (d,e) and tendon ruptures (f).

Function is increasingly disturbed and patients may need help with grooming, dressing and eating.

Extra-articular features These often appear in patients with severe disease. The most characteristic is the appearance of nodules. They are usually found as small subcutaneous lumps, rubbery in consistency, at the back of the elbows, but they also develop in tendons (where they may cause 'triggering' or rupture), in the viscera and the eye. They are pathognomonic of RA, but occur in only 25% of patients.

Less specific features include *muscle wasting*, *lymphadenopathy*, *scleritis*, *nerve entrapment syndromes*, *skin atrophy or ulceration*, *vasculitis* and *peripheral sensory neuropathy*. Marked visceral disease, such as *pulmonary fibrosis*, is rare.

Imaging

X-rays Early on, X-rays show only the features of synovitis: soft-tissue swelling and periarticular osteoporosis. The later stages are marked by the appearance of marginal bony erosions and narrowing of the articular space, especially in the proximal joints of the hands and feet (Figure 3.4). However, most individuals have evidence of erosions within 2 years. In advanced disease, articular destruction and joint deformity are obvious. Flexion and extension views of the cervical spine often show subluxation at the atlantoaxial or mid-cervical levels; surprisingly, this causes few symptoms in the majority of cases.

Ultrasound scanning and MRI The use of other imaging techniques to look at soft-tissue changes and early erosions within joints has become more common. Ultrasound can be particularly useful in defining the presence of synovitis and early erosions. Additional information on vascularity can be obtained if Doppler techniques are used.

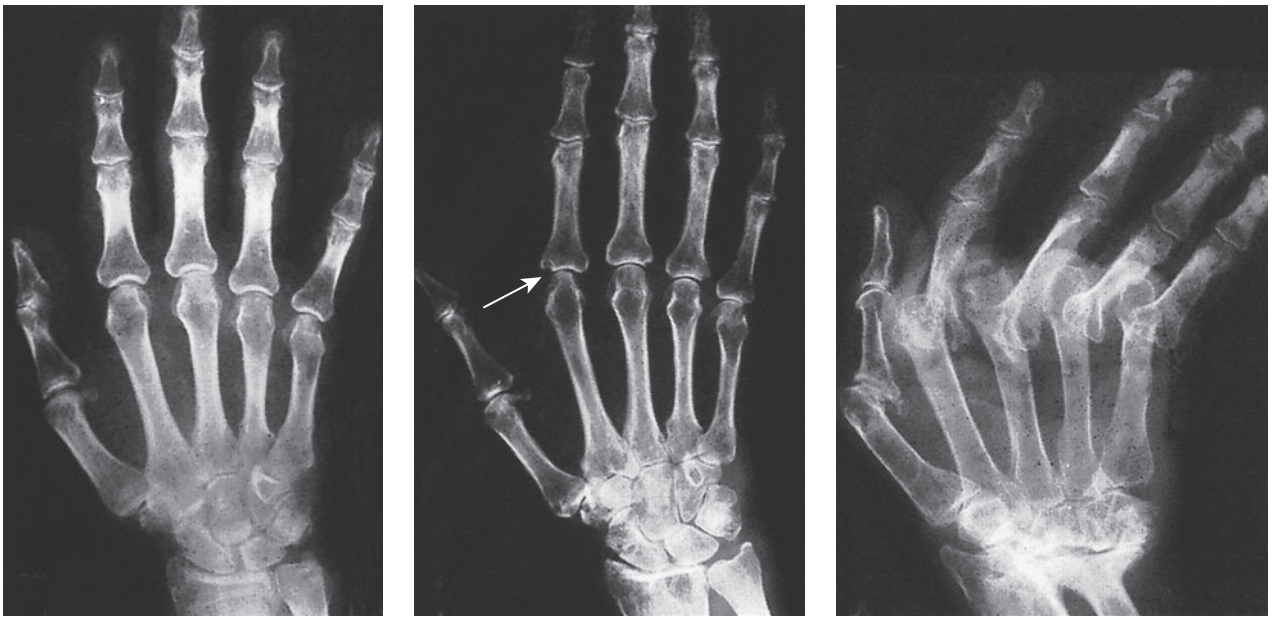
Blood investigations

Normocytic, hypochromic anaemia is common and is a reflection of abnormal erythropoiesis due to disease activity. It may be aggravated by chronic gastrointestinal blood loss caused by non-steroidal anti-inflammatory drugs. In active phases the ESR and CRP concentration are usually raised.

Serological tests for RF are positive in about 80% of patients and antinuclear factors are present in 30%. Neither of these tests is specific and neither is required for a diagnosis of rheumatoid arthritis. Newer tests such as those for anti-CCP antibodies have added much greater specificity but at the expense of sensitivity.

Synovial biopsy

Synovial tissue may be obtained by needle biopsy, via the arthroscope, or by open operation. Unfortunately, most of the histological features of rheumatoid arthritis are non-specific.



(a) (b) (c)

Figure 3.4 Rheumatoid arthritis – X-ray changes The progress of disease is well shown in this patient's X-rays. First, there was only soft-tissue swelling and periarticular osteoporosis; later juxta-articular erosions appeared (arrow); ultimately, the joints became unstable and deformed.

Diagnosis

The usual criteria for diagnosing rheumatoid arthritis are the presence of a bilateral, symmetrical polyarthritis involving the proximal joints of the hands or feet, and persisting for at least 6 weeks. If there are subcutaneous nodules or X-ray signs of periarticular erosions, the diagnosis is certain. *A positive test for RF in the absence of the above features is not sufficient evidence of rheumatoid arthritis, nor does a negative test exclude the diagnosis if the other features are all present.* The chief value of the RF tests is in the assessment of prognosis: persistently high titres herald more serious disease including extra-articular features.

Atypical forms of presentation are not uncommon. The early stages may be punctuated by spells of quiescence, during which the diagnosis is doubted, but sooner or later the more characteristic features appear. Occasionally, in older people, the onset is explosive, with the rapid appearance of severe joint pain and stiffness. Now and then (more so in young women) the disease starts with chronic pain and swelling of a single large joint and it may take months or years before other joints are involved. The presence of tenderness on squeezing across all metacarpophalangeal or metatarsophalangeal joints, early morning stiffness of at least 30 minutes and a raised ESR are highly suggestive of a diagnosis of rheumatoid arthritis. A rapid diagnosis is vital so that early treatment can be started with disease-modifying antirheumatic drugs.

In the differential diagnosis of polyarthritis several disorders must be considered.

Seronegative inflammatory polyarthritis Polyarthritis is a feature of a number of conditions including psoriatic arthritis, adult Still's disease, systemic lupus erythematosus and other connective-tissue diseases. These are considered in later sections.

Ankylosing spondylitis This is primarily an inflammatory disease of the sacroiliac and intervertebral joints, causing back pain and progressive stiffness; however, it may also involve the peripheral joints.

Reiter's disease/reactive arthritis The larger joints and the lumbosacral spine are the main targets. There is usually a history of urethritis or colitis and often also conjunctivitis.

Polyarticular gout Tophaceous gout affecting multiple joints can, at first sight, be mistaken for rheumatoid arthritis. On X-ray, the erosions are quite different from those of rheumatoid arthritis; the diagnosis is clinched by identifying typical birefringent urate crystals in the joint fluid or a nodular tophus.

It is a curious fact that, although both gout and RA are fairly common, the two conditions are rarely seen in the same patient. The reason for this is unknown.

Calcium pyrophosphate deposition disease This condition is usually seen in older people. Typically it affects large joints, but it may occur in the wrist and metacarpophalangeal joints as well. X-ray signs are fairly characteristic and crystals may be identified in synovial fluid or synovium.

Sarcoidosis Sarcoid disease sometimes presents with a symmetrical small-joint polyarthritis and no bone involvement; in other cases a large joint such as the knee or ankle may be involved. Erythema nodosum and hilar lymphadenopathy on chest X-ray are clues to the diagnosis.

Acute sarcoidosis usually subsides spontaneously within 6 months. *Chronic sarcoidosis* produces granulomatous infiltration of lungs, bone, synovium and other organs and is more common in Afro-Caribbean than Caucasian peoples. In addition to polyarthritis and tenosynovitis, there are usually X-ray features of punched-out 'cysts' and cortical erosions in the bones of the hands and feet. The ESR and serum angiotensin converting enzyme (SACE) may be raised. Biopsy of affected tissue shows typical noncaseating granulomas. Treatment with non-steroidal anti-inflammatory drugs (NSAIDs) may be adequate but in more intractable cases corticosteroids or other immunosuppressive therapies are necessary.

Lyme disease This tick-borne spirochaetal infection usually starts with a skin lesion and flu-like symptoms and then spreads to multiple organs. If the initial lesions are missed or left untreated, patients may present with an asymmetrical inflammatory polyarthritis affecting mainly the larger joints. It is most likely to be encountered in known endemic areas in North America, Europe and Asia. In late cases serological tests may be positive. Treatment with doxycycline or one of the newer cephalosporins is usually effective for the arthritic features.

Viral arthritis Viral infections are often associated with a transient polyarthralgia; flu-like illness and a rash will suggest the diagnosis. However, some infections – most typically parvovirus B19 – occasionally cause a symmetrical polysynovitis (including the finger joints) and early morning stiffness, symptoms which may last for several months or may recur over a few years. The absence of 'rheumatoid' X-ray features and subcutaneous nodules will raise suspicions about the diagnosis.

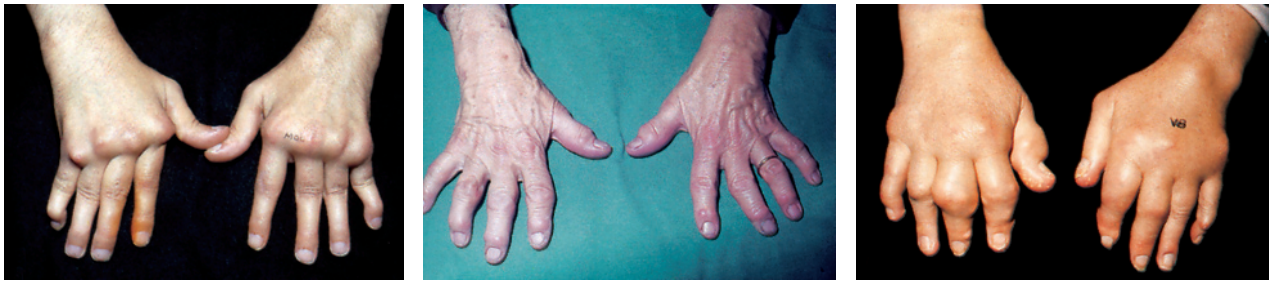
Polymyalgia rheumatica This condition, which is seen mainly in the middle-aged or elderly, is characterized by aching discomfort around the pectoral and pelvic girdles, post-inactivity stiffness and muscular weakness. The joints are not tender but the muscles may be. The ESR and CRP are almost always elevated. Corticosteroids (as little as 10 mg a day) provide rapid and dramatic relief of all symptoms, and this response is often used as a diagnostic test. The condition may be associated with, and certainly carries the risk of, giant cell arteritis which may result in blindness.

Osteoarthritis Polyarticular osteoarthritis (OA), which typically involves the finger joints, is often mistaken for RA. A moment's reflection will usually dispel any doubt: OA always involves the *distal* interphalangeal joints and causes a nodular arthritis with radiologically obvious osteophytes, whereas RA affects the *proximal* joints of the hand and causes predominantly erosive features (see Figure 3.5).

Some confusion may arise from the fact that RA, in its later stages, is associated with loss of articular cartilage and *secondary OA*. Enquiry into the early history will usually untangle the diagnosis. Sometimes, however, RA atypically affects only a few of the larger joints and it is then very difficult to distinguish from OA; X-ray features such as loss of articular cartilage throughout the entire joint and lack of hypertrophic bone changes (sclerosis and osteophytes) should suggest an inflammatory arthritis.

Treatment

There is no cure for rheumatoid arthritis. However, advances in therapy have revolutionized the treatment approach with associated major improvements in outcome. Medical treatment is guided by the principle that inflammation should be reduced rapidly and aggressively. A multidisciplinary approach is helpful from the beginning: ideally the therapeutic team should include a rheumatologist, orthopaedic surgeon, physiotherapist, occupational therapist,



(a) (b) (c)

Figure 3.5 Rheumatoid arthritis – differential diagnosis All three patients presented with painful swollen fingers. In (a) mainly the proximal joints were affected (rheumatoid arthritis); in (b) the distal joints were the worst (Heberden's osteoarthritis); in (c) there were asymmetrical nodular swellings around the joints (gouty tophi).

orthotist and social worker. Their deployment and priorities will vary according to the individual and stage of the disease.

At the onset of the disease both the patient and the doctor will be uncertain about the likely rate of progress. An attempt should be made to determine the likely prognosis. Poor prognosis is associated with female sex, multiple joint involvement, high ESR and CRP, positive RF and anti-CCP, younger age, high BMI, smoking and the presence of erosions at diagnosis.

PRINCIPLES OF MEDICAL MANAGEMENT

Treatment should be aimed at controlling inflammation as rapidly as possible. This is likely to require the use of corticosteroids for their rapid onset (initially oral doses of 30 mg of prednisolone or 120 mg i.m. methylprednisolone may be used). Steroids should be rapidly tapered to prevent significant side effects.

In addition, disease-modifying antirheumatic drugs (DMARDs) should be started at this time. The first choice is now methotrexate at doses of 10–25 mg/week. This may be used initially alone or in combination with sulphasalazine and hydroxychloroquine. Leflunomide can also be considered if methotrexate is not tolerated. Gold and penicillamine are associated with significant side effects and are now used very rarely.

Control of pain and stiffness with non-steroidal anti-inflammatory drugs (NSAIDs) may be needed, maintaining muscle tone and joint mobility by a balanced programme of exercise, and general advice on coping with the activities of daily living.

If there is no satisfactory response to DMARDs, it is wise to progress rapidly to biological therapies such as the TNF inhibitors infliximab, etanercept, golimumab, certolizumab and adalimumab. Other biological therapies include inhibitors of T-cell costimulation (abatacept), IL-6 (tocilizumab) and B-cell depleting therapies (rituximab).

Additional measures include the injection of corticosteroid preparations into inflamed joints and tendon sheaths. It is sometimes feared that such injections may themselves cause damage to articular cartilage or tendons. However, there is little evidence that they are harmful, provided they are used sparingly and with full precautions against infection.

Prolonged rest and immobility is likely to weaken muscles and lead to a worse prognosis. However, some splinting can be helpful at any stage of the disease.

PHYSIOTHERAPY AND OCCUPATIONAL THERAPY

Preventative splinting and orthotic devices may delay the march of events; however, it is important to encourage activity. If these fail to restore and maintain function, operative treatment is indicated.

SURGICAL MANAGEMENT

At first this consists mainly of soft-tissue procedures (synovectomy, tendon repair or replacement and joint stabilization); in some cases osteotomy may be more appropriate.

In late rheumatoid disease, severe joint destruction, fixed deformity and loss of function are clear indications for reconstructive surgery. Arthrodesis, osteotomy and arthroplasty all have their place and are considered in the appropriate chapters. However, it should be recognized that patients who are no longer suffering the pain of active synovitis and who are contented with a limited pattern of life may not want or need heroic surgery merely to improve their anatomy. Careful assessment for occupational therapy, the provision of mechanical aids and adjustments to their home environment may be much more useful. It appears safe to continue methotrexate during elective orthopaedic surgery. However, doses of corticosteroids should be as low as possible and biological therapies such as the TNF inhibitors should be stopped prior to surgery where possible.

BOX 3.1 KEY ELEMENTS IN MEDICAL TREATMENT

Identify patients with RA as early as possible.

Start disease-modifying antirheumatic drugs (DMARDs) immediately.

Consider combination therapy with multiple DMARDs.

If DMARDs fail, progress rapidly to biological therapies such as the TNF inhibitors.

Complications

Fixed deformities The perils of rheumatoid arthritis are often the commonplace ones resulting from ignorance and neglect. Early assessment and planning should prevent postural deformities, which will result in joint contractures.

Muscle weakness Even mild degrees of myopathy or neuropathy, when combined with prolonged inactivity, may lead to profound muscle wasting and weakness. This should be prevented by control of inflammation, physiotherapy and pain control, if possible; if not, the surgeon must be forewarned of the difficulty of postoperative rehabilitation.

Joint rupture Occasionally the joint capsule ruptures and synovial contents spill into the soft tissues. Treatment is directed at the underlying synovitis, i.e. splintage and injection of the joint, with synovectomy as a second resort.

Infection Patients with rheumatoid arthritis – and even more so those on corticosteroid therapy – are susceptible to infection. Sudden clinical deterioration, or increased pain in a single joint, should alert one to the possibility of septic arthritis and the need for joint aspiration.

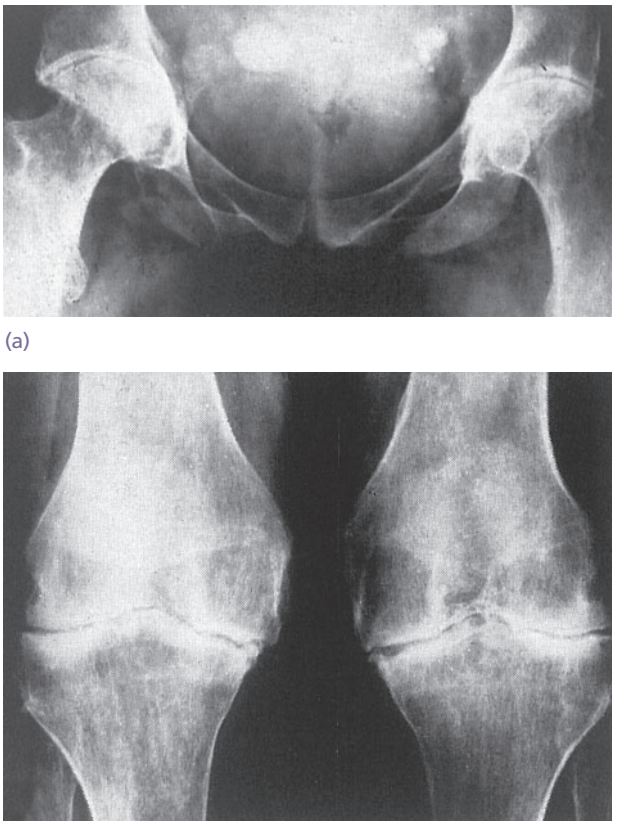
Spinal cord compression This is a rare complication of cervical spine (atlantoaxial) instability. The onset of weakness and upper motor neuron signs in the lower limbs is suspicious. If they occur, immobilization of the neck is essential and spinal fusion should be carried out as soon as possible.

Systemic vasculitis Vasculitis is a rare but potentially serious complication. Corticosteroids and immunosuppressives such as intravenous cyclophosphamide may be required.

Amyloidosis This is another rare but potentially lethal complication of long-standing rheumatoid arthritis. The patient presents with proteinuria and progressive renal failure. Finding amyloid in a rectal or renal biopsy makes the diagnosis. Aggressive control of inflammation has reduced this complication significantly.

Prognosis

Rheumatoid arthritis runs a variable course. When the patient is first seen, it is difficult to predict the outcome, but high titres of RF and anti-CCP, periarticular erosions, rheumatoid nodules, severe muscle wasting, joint contractures and evidence of vasculitis are bad prognostic signs. Women, on the whole, fare somewhat worse than men. Without effective treatment about 10% of patients improve steadily after the first attack of active synovitis; 60% have intermittent phases of disease activity and remission, but with a slow downhill course over many years; 20% have severe joint erosion, which is usually evident within the first 5 years (Figure 3.6); and 10% end up completely disabled. In addition, a reduction in life expectancy by 5–10 years is common and is often due to premature ischaemic heart disease. However, early aggressive medical treatment appears to reduce the morbidity and mortality.



(a)

(b)

Figure 3.6 Rheumatoid arthritis – aftermath After the acute inflammatory phase has passed, the patient may be left with features of secondary osteoarthritis, especially in the hips (a) and the knees (b).

AXIAL SPONDYLOARTHROPATHIES INCLUDING ANKYLOSING SPONDYLITIS

These are generalized chronic inflammatory diseases, but their effects are seen mainly in the spine and sacroiliac joints. Definitions have changed in the last decade to reflect the fact that ankylosing spondylitis (AS) is the end stage of a disease process best described by the term axial spondyloarthropathy (axial SPA). AS is characterized by pain and stiffness of the back, with variable involvement of the hips and shoulders and (more rarely) the peripheral joints. Its reported prevalence is 0.1–0.2% in Western Europe and North America, but it is much lower in Japanese and African peoples. Males are affected more frequently than females (estimates vary from 2:1 to 10:1) and the usual age at onset is between 15 and 25 years. There is a strong tendency to familial aggregation and association with the genetic markers HLA-B27 and ERAP1.

Cause

There is considerable evidence for regarding ankylosing spondylitis (AS) as a genetically determined immunopathological disorder. The disease is much more common in family members of patients than in the general population – HLA-B27 is present in over 95% of Caucasian patients and in half of their first-degree relatives (as compared with 8% of the general population); and racial groups with an unusually low prevalence of AS also show a very low prevalence of HLA-B27 (e.g. less than 1% in Japanese people).

There are various theories about the 'triggering factor' that initiates the abnormal immune response. It may be a bacterial antigen, which closely resembles HLA-B27 that induces an antibody response, which also targets the HLA-B27 positive cells; or, as in the case of RA, the HLA-B27 molecule may be involved in the presentation of a specific antigen to the T-cells, which then react with the antigen-presenting cells. Since classic ankylosing spondylitis is sometimes associated with genitourinary or bowel infection, and disorders such as Reiter's disease and ulcerative colitis cause vertebral and sacroiliac changes indistinguishable from those of ankylosing spondylitis, it has been suggested that the putative organism may be carried to the spine by local lymphatic drainage.

Pathology

There are two basic lesions: synovitis of diarthrodial joints and inflammation at the fibro-osseous junctions of syndesmotic joints and tendons. The preferential involvement of the insertion of tendons and ligaments (the entheses) has resulted in the term *enthesopathy*.

Synovitis of the sacroiliac and vertebral facet joints causes destruction of articular cartilage and periarticular bone. The costovertebral joints also are frequently involved, leading to diminished respiratory excursion. When peripheral joints are affected, the same changes occur.

Inflammation of the fibro-osseous junctions affects the intervertebral discs, sacroiliac ligaments, symphysis pubis, manubrium sterni and the bony insertions of large tendons. Pathological changes proceed in three stages: (1) an inflammatory reaction with cell infiltration, granulation tissue formation and erosion of adjacent bone; (2) replacement of the granulation tissue by fibrous tissue; and (3) ossification of the fibrous tissue, leading to ankylosis of the joint.

Ossification across the surface of the disc gives rise to small bony bridges or syndesmophytes linking adjacent vertebral bodies. If many vertebrae are involved the spine may become absolutely rigid.

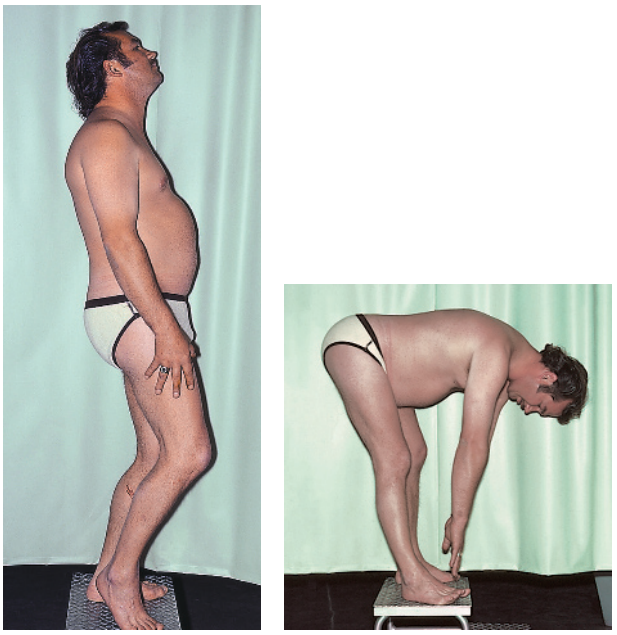
Clinical features

The disease starts insidiously: a teenager or young adult complains of backache and stiffness recurring at intervals over a number of years. This is often diagnosed as 'simple mechanical back pain', but the symptoms are worse in the early morning and after inactivity. Referred pain in the buttocks and thighs may appear as 'sciatica' and some patients are mistakenly treated for intervertebral disc prolapse. Gradually pain and stiffness become continuous and other symptoms begin to appear: general fatigue, pain and swelling of joints, tenderness at the insertion of the Achilles tendon, 'foot strain', or intercostal pain and tenderness.

Occasionally the disease starts with pain and slight swelling in a peripheral joint such as the ankle, or pain and stiffness of the hip. Sooner or later, though, backache will come to the fore. In women the axial skeletal disease may remain restricted to the sacroiliac joints making diagnosis challenging.

Early on there is little to see apart from slight flattening of the lower back and limitation of extension in the lumbar spine (Figure 3.7). There may be diffuse tenderness over the spine and sacroiliac joints, or (occasionally) swelling and tenderness of a single large joint.

In established cases the posture is typical: loss of the normal lumbar lordosis, increased thoracic kyphosis and a forward thrust of the neck; upright posture and balance are maintained by standing with the hips and knees slightly flexed, and in late cases these may become fixed deformities. Spinal movements are diminished in all directions, but loss of extension is always the earliest and the most severe disability. It is revealed dramatically by the 'wall test': the patient is asked to stand with his back to the wall; heels, buttocks, scapulae and occiput should all be able to touch the wall simultaneously. If extension is seriously diminished, the patient will find this impossible. In the most advanced stage the spine may be completely ankylosed from occiput to sacrum – sometimes in positions of grotesque deformity. Marked loss of cervical extension may restrict the line of vision to a few paces.



(a) (b)

Figure 3.7 Ankylosing spondylitis – early The cardinal clinical feature is marked stiffness of the spine. (a) This patient manages to stand upright by keeping his knees slightly flexed. (b) It looks as if he can bend down to touch his toes, but his back is rigid and all the movement takes place at his hips.

Chest expansion, which should be at least 7 cm in young men, is often markedly decreased. In old people, who may have pulmonary disease, this test is unreliable.

Peripheral joints (usually shoulders, hips and knees) are involved in over a third of the patients; they show the features of inflammatory arthritis – swelling, tenderness, effusion and loss of mobility. There may also be tenderness of the ligament and tendon insertions close to a large joint or under the heel.

Extraskeletal manifestations General fatigue and loss of weight are common. Acute anterior uveitis occurs in about 25% of patients; it usually responds well to treatment but, if neglected, may lead to permanent damage including glaucoma. Other extraskeletal disorders, such as aortic valve disease, carditis and pulmonary fibrosis (apical), are rare and occur very late in the disease.

Imaging

X-rays The cardinal sign – and often the earliest – is erosion and fuzziness of the sacroiliac joints. Later there may be periarticular sclerosis, especially on the iliac side of the joint and finally bony ankylosis.

The earliest vertebral change is flattening of the normal anterior concavity of the vertebral body ('squaring'). Later, ossification of the ligaments around the intervertebral discs produces delicate bridges (syndesmophytes) between adjacent vertebrae (Figure 3.8). Bridging at several levels gives the appearance of a 'bamboo spine'.

Osteoporosis is common in long-standing cases and there may be hyperkyphosis of the thoracic spine due to wedging of the vertebral bodies.

Peripheral joints may show erosive arthritis or progressive bony ankylosis.

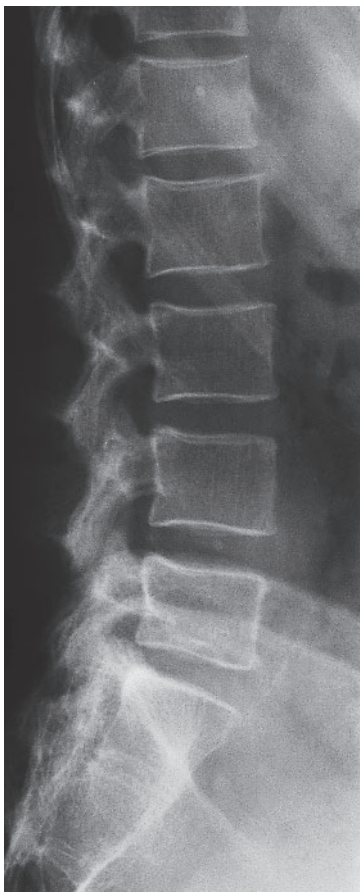
MRI MRI allows detailed investigation of sacroiliac joints and may show typical erosions and features of inflammation such as bone oedema. Various techniques including gadolinium contrast can be used to demonstrate inflammatory lesions in other areas of the spine.

Special investigations

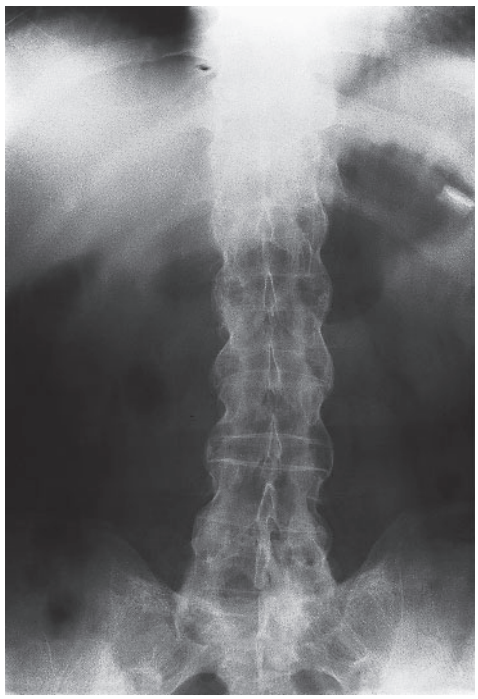
The ESR and CRP are usually elevated during active phases of the disease. HLA-B27 is present in 95% of cases. Serological tests for rheumatoid factor are usually negative.

Diagnosis

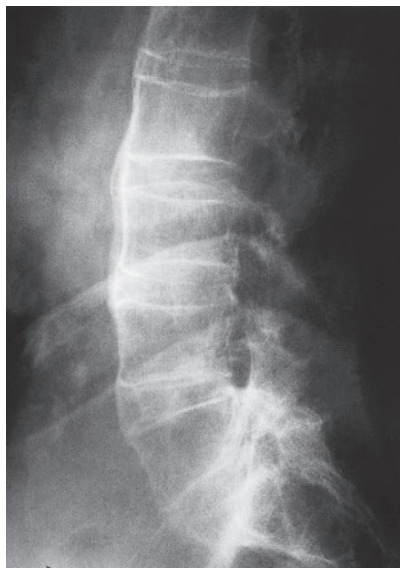
Diagnosis is easy in patients with spinal rigidity and typical deformities, but it is often missed in those with early disease before radiographic changes are



(a)



(b)



(c)

Figure 3.8 Ankylosing spondylitis – X-rays (a) An early sign is 'squaring' of the lumbar vertebrae. (b,c) Bony bridges (syndesmophytes) between the vertebral bodies convert the spine into a rigid column.

seen (non-radiographic axial SPA) or unusual forms of presentation. In over 10% of cases the disease starts with an asymmetrical inflammatory arthritis – usually of the hip, knee or ankle – and it may be several years before back pain appears. Atypical onset is more common in women, who may show less obvious changes in the sacroiliac joints. A history of AS in a close relative is strongly suggestive.

Mechanical disorders Low back pain in young adults is usually attributed to one of the more common disorders such as muscular strain, facet joint dysfunction or spondylolisthesis. These conditions differ from AS in several ways: the onset of pain is related to specific physical activities, stiffness is less pronounced and symptoms are eased rather than aggravated by inactivity. Tenderness is also more localized and the peripheral joints are normal.

Diffuse idiopathic hyperostosis (Forestier's disease) This is a fairly common disorder, predominantly of older men, characterized by widespread ossification of ligaments and tendon insertions. X-rays show pronounced but asymmetrical intervertebral spur formation and bridging throughout the dorsolumbar spine. Although it bears a superficial resemblance to AS, it is not an inflammatory disease, spinal pain and stiffness are seldom severe, the sacroiliac joints are not eroded and the ESR is normal.

Other seronegative spondyloarthropathies A number of disorders are associated with vertebral and sacroiliac lesions indistinguishable from those of ankylosing spondylitis. They are *Reiter's disease*, *psoriatic arthritis*, *ulcerative colitis*, *Crohn's disease*, *Whipple's disease* and *Behçet's syndrome*. In each there are certain characteristic features: the rash or nail changes of psoriasis, intestinal ulceration in inflammatory bowel disease, genitourinary and ocular inflammation in Reiter's disease, buccal and genital ulceration in Behçet's syndrome. Yet there is considerable overlap between them: all show some familial aggregation and many are associated with the histocompatibility antigen, HLAB27. Patients with one of these disorders (including AS) often have close relatives with another, or with a positive HLA-B27.

Treatment

The disease can be as damaging to a patient as rheumatoid arthritis but some continue to lead an active life. Treatment consists of: (1) general measures to maintain satisfactory posture and preserve movement; (2) anti-inflammatory drugs to counteract pain and stiffness; (3) the use of TNF inhibitors for severe disease (with inhibitors of other cytokines such as IL-17 and IL-12/23 in development); and (4) operations to correct deformity or restore mobility.

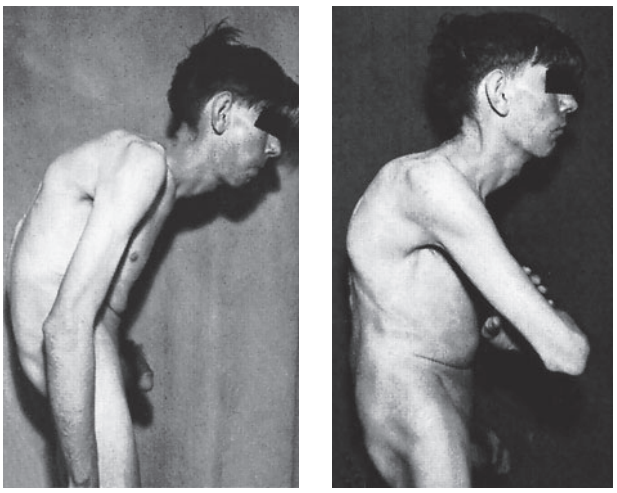
General measures Patients are encouraged to remain active and follow their normal pursuits. They should be taught how to maintain satisfactory posture and urged to perform spinal extension exercises every day. Swimming, dancing and gymnastics are ideal forms of recreation. Rest and immobilization are contraindicated because they tend to increase the general feeling of stiffness.

Non-steroidal anti-inflammatory drugs It is doubtful whether these drugs prevent or retard the progress to ankylosis, but they do control pain and counteract soft-tissue stiffness, thus making it possible to benefit from exercise and activity. They may have to be continued for many years.

TNF inhibitors With the introduction of the TNF inhibitors it has become possible to treat the underlying inflammatory processes active in AS. This can result in significant improvement in disease activity including remission. These therapies are generally reserved for individuals who have failed to be controlled with non-steroidal anti-inflammatory drugs.

Operation Significantly damaged hips can be treated by joint replacement, though this seldom provides more than moderate mobility. Moreover, the incidence of infection is higher than usual and patients may need prolonged rehabilitation.

Deformity of the spine may be severe enough to warrant lumbar or cervical osteotomy (Figure 3.9). These are difficult and potentially hazardous procedures; fortunately, with improved activity and exercise



(a) (b)

Figure 3.9 Ankylosing spondylitis – operative treatment Spinal osteotomy is occasionally performed to correct a severe, rigid deformity. (a) Before operation this man could see only a few paces ahead; (b) after osteotomy his back is still rigid but his posture, function and outlook are improved.

programmes, they are seldom needed. If spinal deformity is combined with hip stiffness, hip replacements (permitting full extension) often suffice.

Complications

Spinal fractures The spine is often both rigid and osteoporotic; fractures may be caused by comparatively mild injuries. The commonest site is C5–7, but it is prudent to X-ray the entire spine in accident victims who have AS. Treatment in these cases is directed at preventing further deformity.

Hyperkyphosis In long-standing cases the spine may become severely kyphotic, so much so that the patient has difficulty lifting his head to see in front of his feet.

Spinal cord compression This is uncommon, but it should be thought of in patients who develop long tract symptoms and signs. It may be caused by atlantoaxial subluxation or by ossification of the posterior longitudinal ligament.

Lumbosacral nerve root compression Patients may occasionally develop root symptoms, including lower limb weakness and paraesthesia, in addition to their 'usual' pelvic girdle symptoms.

PERIPHERAL SPONDYLOARTHROPATHIES

Reiter's syndrome and reactive arthritis

The syndrome described by Hans Reiter in 1916 (and 100 years before that by Benjamin Brodie) is a clinical triad of *urethritis*, *arthritis* and *conjunctivitis* occurring some weeks after either *dysentery* or *genitourinary infection* (Figure 3.10). It is now recognized that this is one of the classic forms of reactive arthritis, i.e. an aseptic inflammatory arthritis associated with non-specific infection (often urogenital or bowel).

Its prevalence is difficult to assess, but it is probably the commonest type of large-joint polyarthritis in young men. It is thought to occur in 1–3% of all people who develop either non-specific urogenital infection or *Shigella* dysentery, but its incidence may be as high as 25% in those who are HLA-B27 positive. Men are affected more often than women (the ratio is about 10:1), but this may simply reflect the difficulty of diagnosing the genitourinary infection in women. The usual age at onset is between 20 and 40 years, but children are affected too – perhaps after an episode of diarrhoea.

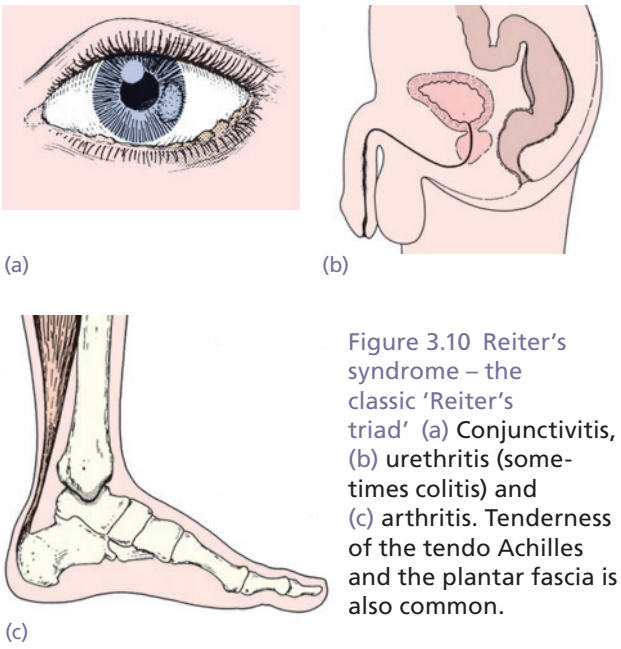


Figure 3.10 Reiter's syndrome – the classic 'Reiter's triad' (a) Conjunctivitis, (b) urethritis (sometimes colitis) and (c) arthritis. Tenderness of the tendo Achilles and the plantar fascia is also common.

Cause

Familial aggregation, overlap with other forms of seronegative spondyloarthropathy in first-degree relatives and a close association with HLA-B27 point to a genetic predisposition, the bowel or genitourinary infection acting as a trigger. Gut pathogens include *Shigella flexneri*, *Salmonella*, *Campylobacter* species and *Yersinia enterocolitica*. *Lymphogranuloma venereum* and *Chlamydia trachomatis* have been implicated as sexually transmitted infections. All these bacteria can survive in human cells; assuming that either the bacterium or a peptide bacterial fragment acts as the antigen, the pathogenesis could be the same as that suggested for ankylosing spondylitis.

Pathology

The pathological changes are essentially the same as those in ankylosing spondylitis, with the emphasis first on subacute large-joint synovitis and in some individuals with a chronic disease course tending towards sacroiliitis and spondylitis.

Clinical features

Acute phase The acute phase of the disease is marked by an asymmetrical inflammatory arthritis of the lower limb joints – usually the knee and ankle but often the tarsal and toe joints as well. The joint may be acutely painful, hot and swollen with a tense effusion, suggesting gout or infection. Tendo Achilles tenderness and plantar fasciitis (evidence of enthesopathy) are common, and the patient may complain of backache even in the early stage. Conjunctivitis, urethritis and bowel infections are often mild and easily

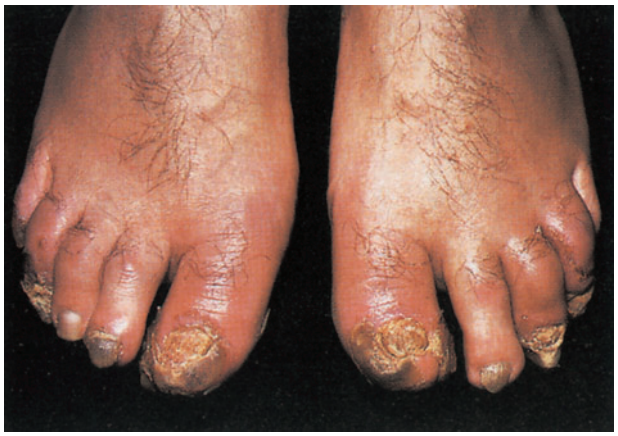


Figure 3.11 Reiter's disease – other features The characteristic pustular dermatitis of the feet – keratoderma blennorrhagicum.

missed; the patient should be carefully questioned about symptoms during the previous few weeks. Cystitis and cervicitis may occur in women.

Less frequent, but equally characteristic, features are a vesicular or pustular dermatitis of the feet (keratoderma blennorrhagica – Figure 3.11), balanitis and mild buccal ulceration.

The acute disorder usually lasts for a few weeks or months and then subsides, but most patients have either recurrent attacks of arthritis or other features of chronic disease.

Chronic phase The chronic phase is more characteristic of a spondyloarthropathy. Over half of the patients with Reiter's disease complain of mild, recurrent episodes of polyarthritis (including upper limb joints). About half of those again develop sacroiliitis and spondylitis with features resembling those of ankylosing spondylitis. Uveitis is also fairly common and may give rise to posterior synechiae and glaucoma.

X-rays

Sacroiliac and vertebral changes are similar to those of ankylosing spondylitis. If peripheral joints are involved, they may show features of erosive arthritis.

Special investigations

Tests for HLA-B27 are positive in 75% of patients with sacroiliitis. The ESR may be high in the active phase of the disease. The causative organism can sometimes be isolated from urethral fluids or faeces, and tests for antibodies may be positive.

Diagnosis

The diagnosis should be considered in any young adult who presents with an acute or subacute arthritis in the lower limbs. It is more likely to be missed in women, in children and in those with very mild (and often forgotten) episodes of genitourinary or bowel infection. Some patients never develop the full syndrome and one should be alert to the *formes fruste* with large-joint arthritis alone.

Gout and infective arthritis Reiter's disease, gout and infection should all be considered in the differential diagnosis of inflammation in a large peripheral joint. Examination of synovial fluid for organisms and crystals may provide important clues.

Gonococcal arthritis Gonococcal arthritis takes two forms: (1) bacterial infection of the joint; and (2) a reactive arthritis with sterile joint fluid. A history of genitourinary infection further complicates the distinction from Reiter's disease, and diagnosis may depend on identifying the organism or gonococcal antibodies.

Enteropathic arthritis Ulcerative colitis and Crohn's disease may be associated with subacute synovitis, causing pain and swelling of one or more of the peripheral joints. These subside when the intestinal disease is controlled.

Treatment

Initial treatment for Reiter's disease should be aimed at ensuring the infectious organism responsible has been cleared. This is particularly important for sexually transmitted infections such as *Chlamydia trachomatis*.

Even if the triggering infection is identified, treating it will have no effect on the reactive arthritis. However, there is some evidence that treatment of *Chlamydia* infection with tetracycline for periods of up to 3 months can reduce the risk of recurrent joint disease.

Symptomatic treatment could include the use of analgesia and non-steroidal anti-inflammatory drugs. If the inflammatory response is aggressive, local injection of corticosteroids or even intramuscular methylprednisolone may be useful. If symptoms and signs do not resolve, DMARDs used in the treatment of RA may be needed. Topical steroids may be used for uveitis.

Psoriatic arthritis

Psoriatic arthritis is characterized by seronegative polysynovitis, erosive (sometimes very destructive) arthritis, enthesitis and dactylitis and a significant incidence of sacroiliitis and spondylitis.

The prevalence of psoriasis is 1–2%, but only about 5% of those affected will develop psoriatic arthritis. The usual age at onset is 30–50 years (often later than the skin lesions).

Cause

As with the other spondyloarthropathies, there is a strong genetic component: patients often give a family history of psoriasis; there is a significantly increased incidence of other spondyloarthropathies in close relatives; and 60% of those with psoriatic spondylitis or sacroiliitis have HLA-B27.

Psoriatic skin lesions may well be a reactive phenomenon, and the joint lesions a form of 'reactive arthritis'. However, no specific trigger agent has thus far been identified.

Pathology

The joint changes are similar to those in rheumatoid arthritis – chronic synovitis with cell infiltration and exudate, going on to fibrosis. Cartilage and bone destruction may be unusually severe ('arthritis mutilans'). However, rheumatoid nodules are not seen. Sacroiliac and spine changes, which occur in about 30% of patients, are similar to those in ankylosing spondylitis.

Clinical features

The patient may present with one of several patterns of joint involvement. These include: arthritis of distal interphalangeal joints (Figure 3.12), 'arthritis mutilans', asymmetrical large joint oligoarthritis and patterns mimicking rheumatoid arthritis or ankylosing spondylitis. Psoriasis of the skin or nails usually precedes the arthritis, but hidden lesions (in the natal cleft or umbilicus) are easily overlooked.

The condition can progress slowly or very rapidly and may become quiescent. Sometimes (particularly in women) joint involvement is more symmetrical, and in these cases the condition may be indistinguishable from seronegative rheumatoid arthritis. Asymmetrical swelling of two or three fingers may be due to a combination of interphalangeal arthritis and tenosynovitis.

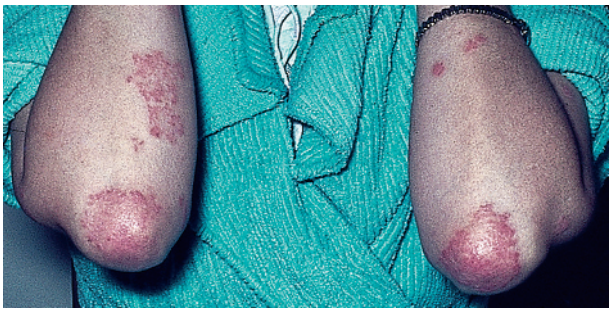
Sacroiliitis and spondylitis are seen in about one-third of patients, and occasionally this is the predominant change with a clinical picture resembling ankylosing spondylitis. As in the other spondyloarthropathies, heel pain (*enthesitis*) is not uncommon.

In the worst cases both the spine and the peripheral joints may be involved. Fingers and toes are severely deformed due to erosion and instability of the interphalangeal joints (*arthritis mutilans* – Figure 3.13).

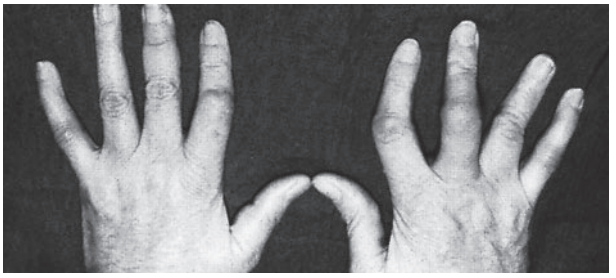
Ocular inflammation occurs in about 30% of patients.

Imaging

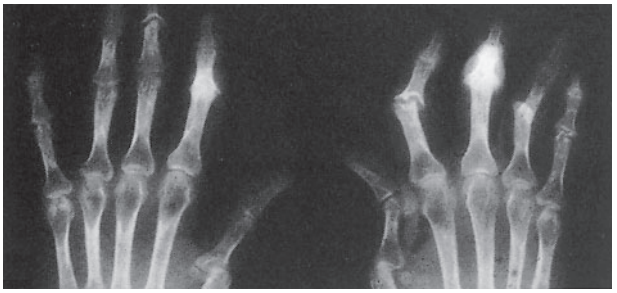
X-ray examination may show severe destruction of the interphalangeal joints of the hands and feet; changes



(a)

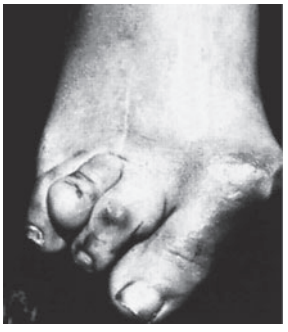


(b)

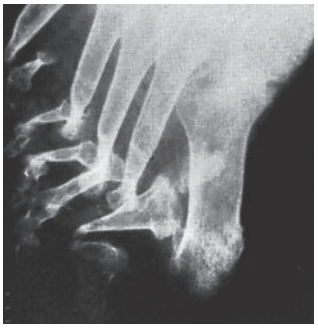


(c)

Figure 3.12 Psoriatic arthritis (1) (a) Psoriasis of the elbows and forearms; (b) typical finger deformities and (c) X-rays show distal joint involvement – clearly the disease is not simply rheumatoid arthritis in a patient with psoriasis.



(a)



(b)

Figure 3.13 Psoriatic arthritis (2) (a and b) The feet and toes are often involved. In this case the patient developed a severely destructive form of the disease (arthritis mutilans).

in the large joints are similar to those of rheumatoid disease. Sacroiliac erosion is fairly common; if the spine is involved the appearances are identical to those of ankylosing spondylitis.

Ultrasound scanning and *MRI* may show greater definition of the extent and activity of synovitis.

Special investigations

Tests for rheumatoid factor are almost always negative. HLA-B27 occurs in 50–60%, especially in those with overt sacroiliitis.

Diagnosis

The main difficulty is to distinguish 'psoriatic arthritis' from 'psoriasis with seronegative RA'. The important distinguishing features of psoriatic arthritis are: (1) asymmetrical joint distribution; (2) involvement of distal finger joints; (3) the presence of sacroiliitis or spondylitis; and (4) the absence of rheumatoid nodules.

Treatment

In mild disease no more than topical preparations to control the skin disease and NSAIDs for the arthritis are needed. In resistant forms of arthritis, immunosuppressive agents (methotrexate) and TNF inhibitors have proved effective. Inhibitors of other cytokines such as IL-17 and IL-12/23 are in development and oral therapies that target intracellular signalling are also available (apremilast). Surgery may be needed for unstable joints. Arthrodesis of the distal interphalangeal joints may greatly improve function.

Enteropathic arthritis

Both Crohn's disease and ulcerative colitis may be associated with either peripheral arthritis or sacroiliitis and spondylitis.

Peripheral arthritis

Peripheral arthritis is fairly common, occurring in about 15% of patients with inflammatory bowel disease. Typically one or perhaps a few of the larger joints are involved. Pain and swelling may appear quite suddenly and last for 2–3 months before subsiding. Synovitis is usually the only feature but joint erosion can occur. Men and women are affected with equal frequency and there is no particular association with HLA-B27.

Treatment is directed at the underlying disorder: attacks of arthritis are often triggered by a flare-up of bowel disease and when the latter is brought under control the arthritis can disappear. Anti-inflammatory drugs should not generally be used as they may have a deleterious effect on the bowel disease. Other treatment options are local corticosteroid injection and disease-modifying treatments such as methotrexate. This may also improve the bowel disease. In severe cases TNF inhibitors may be needed.

Sacroiliitis and spondylitis

This pattern is seen in about 10% of patients with inflammatory bowel disease, and in half of these patients the clinical picture closely resembles that of ankylosing spondylitis. HLA-B27 is positive in 60% and there is an increased incidence of ankylosing spondylitis in close relatives. Unlike the peripheral arthritis, sacroiliitis shows no temporal relationship to gastrointestinal inflammation and its course is unaffected by treatment of the bowel disease. Management is the same as that of ankylosing spondylitis.

Complications

In addition to spondyloarthritis, there are several unusual but important complications of inflammatory bowel disease that may confuse the clinical picture.

Septic arthritis of the hip Infection may spread directly from the bowel. The patient presents with a fever and pain in the groin. Hip movements are limited and there may be swelling due to an abscess. Treatment is by antibiotics and operative drainage.

Psoas abscess In Crohn's disease a posterior fistula may track into the psoas sheath. The patient complains of back pain and may develop a typical psoas abscess with pain in the hip, limitation of movement and a tender mass in the groin. Treatment is by operative drainage of the abscess.

Osteopaenia Patients with chronic bowel disease often develop osteoporosis and osteomalacia – partly due to malabsorption and partly as a consequence of treatment with corticosteroids. Compression fractures of the spine may cause severe back pain.

Juvenile idiopathic arthritis

Juvenile idiopathic arthritis (JIA) is the preferred term for non-infective inflammatory joint disease of more than 3 months' duration in children under 16 years of age. It embraces a group of disorders in all of which pain, swelling and stiffness of the joints are common features. The prevalence is about 1 per 1000 children, and boys and girls are affected with equal frequency.

The cause is similar to that of rheumatoid arthritis: an abnormal immune response to some antigen in children with a particular genetic predisposition. However, rheumatoid factor is usually absent.