



CHRONIC HAEMOPHILIC ARTHROPATHY

A/PROFESSOR JOHN YORK
INTERNATIONAL HAEMOPHILIA TRAINING
CENTRE
ROYAL PRINCE ALFRED HOSPITAL
SYDNEY

HAEMOPHILIA

SEVERITY

FREQUENCY OF HAEMARTHROSES

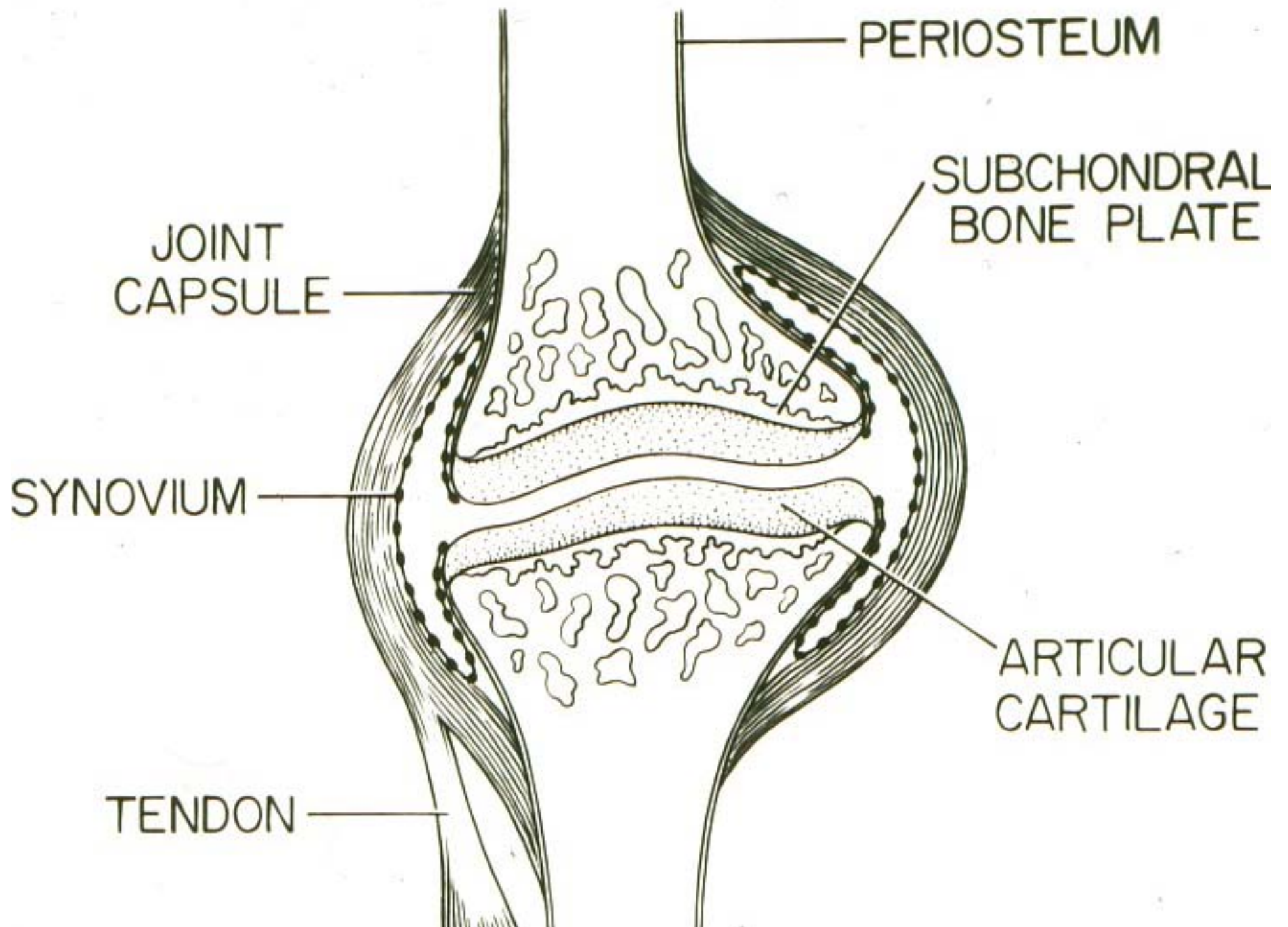
FACTOR LEVEL (%NORMAL)

JOINT INVOLVED

- 1 SEVERE (50% of cases)
- 1-5 MODERATELY SEVERE
- 5-20 MODERATE
- 20-60 MILD



KNEE
ELBOW
ANKLE
SHOULDER
HIP
OTHER



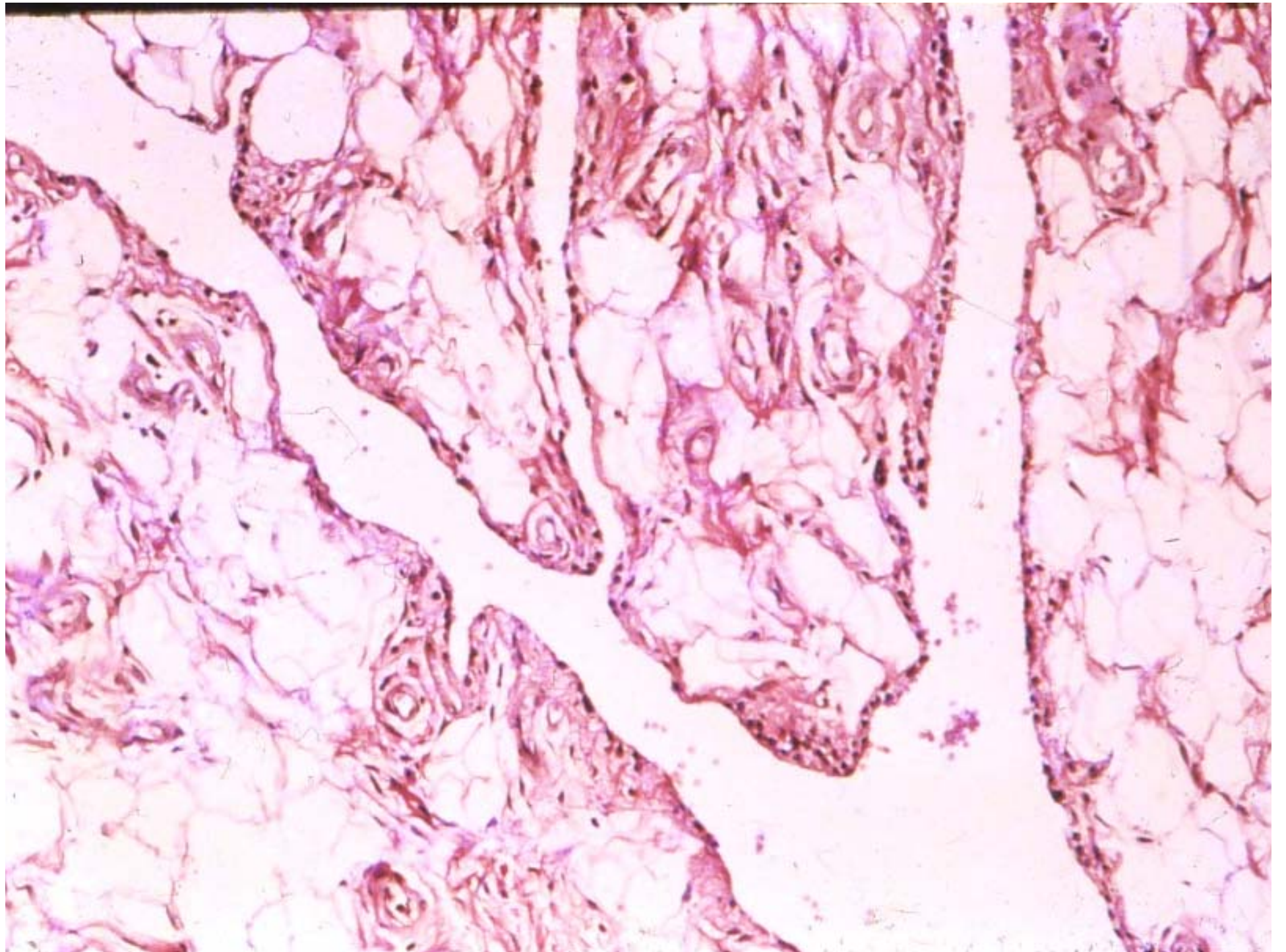
HAEMOPHILIC ARTHROPATHY

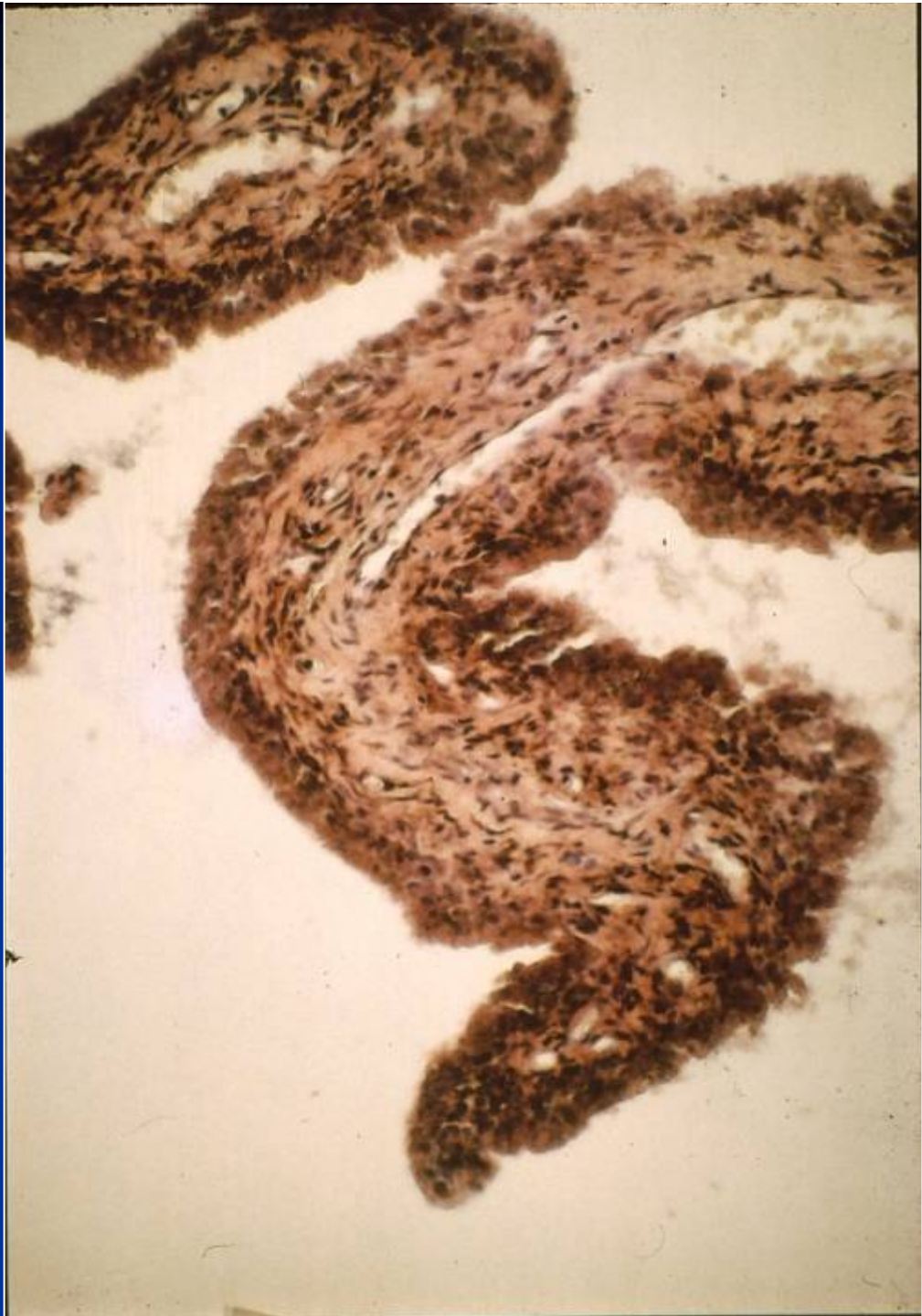
ACUTE HAEMARTHROSIS

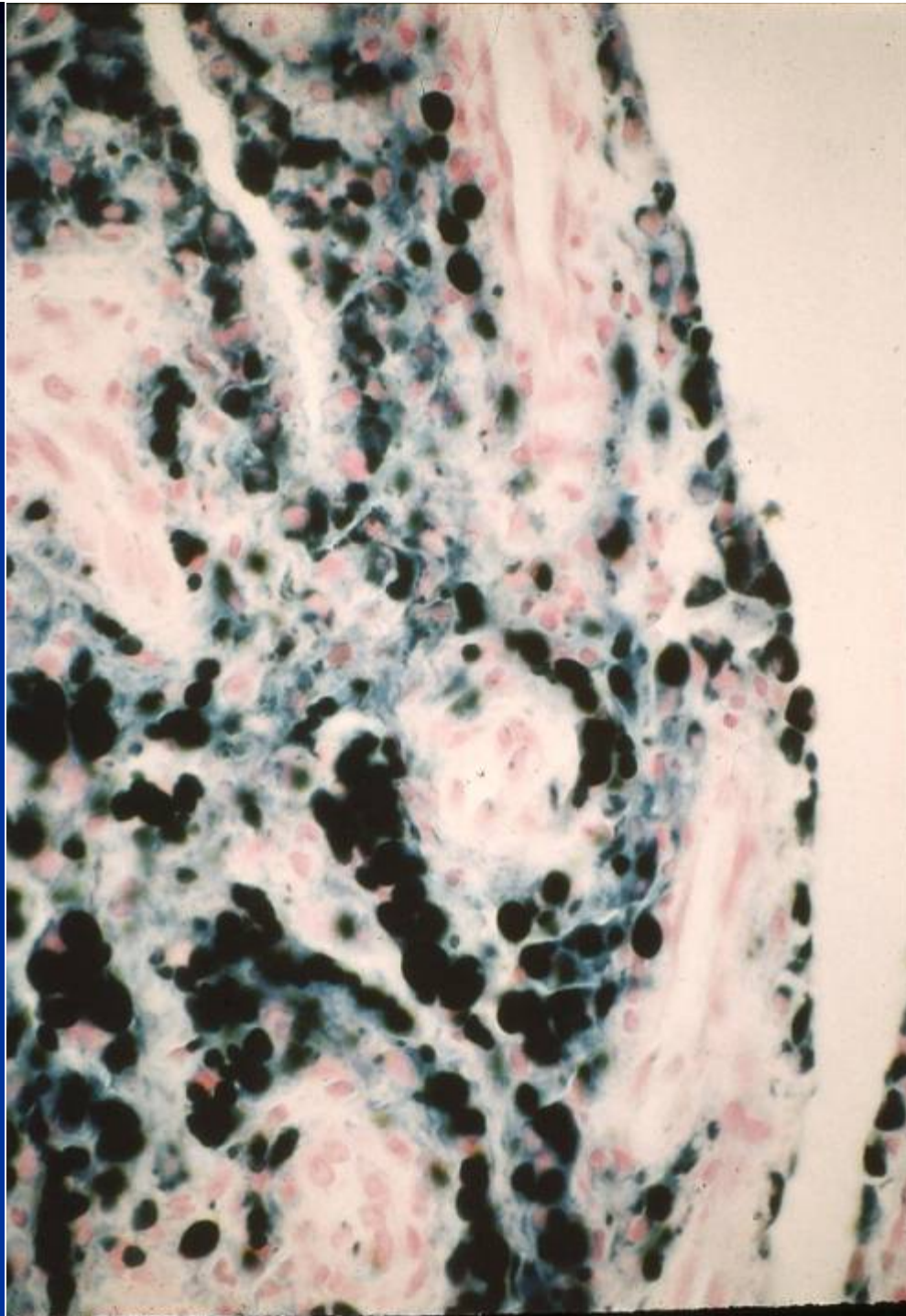
- PRODROMAL STIFFNESS AND PAIN.
TENSE, SWOLLEN, HARD, HOT, TENDER.
SHINY RED SKIN.
- KNEES
ELBOWS
ANKLES
HIPS
SPINE - RARELY
HAND - MAINLY POST TRAUMA
- RAPID PAIN RELIEF
WITH APPROPRIATE FACTOR REPLACEMENT

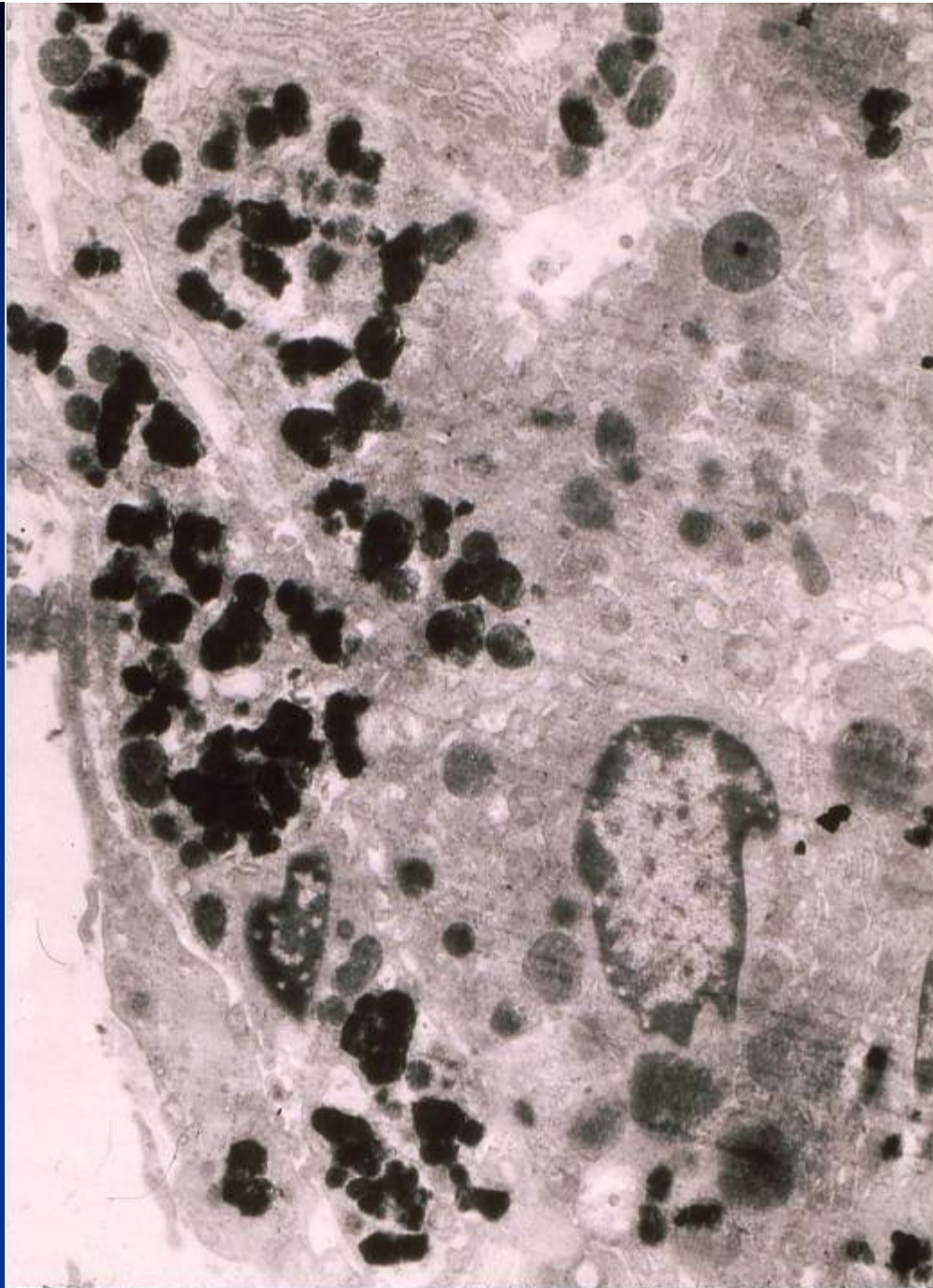
PATHOLOGY

HISTOLOGY









CLIN. PATHOLOGY

- ACUTE BLEEDS
- SUB-ACUTE ARTHROPATHY
- CHRONIC ARTHROPATHY



SUB ACUTE HAEMARTHROSIS

- USUALLY AFTER 2 OR MORE ACUTE BLEEDS.
- PERSISTS DESPITE TREATMENT.
- THICK BOGGY SYNOVIUM.
- MODERATE RESTRICTION OF JOINT MOTION.
- PAIN NOT PROMINENT FEATURE.





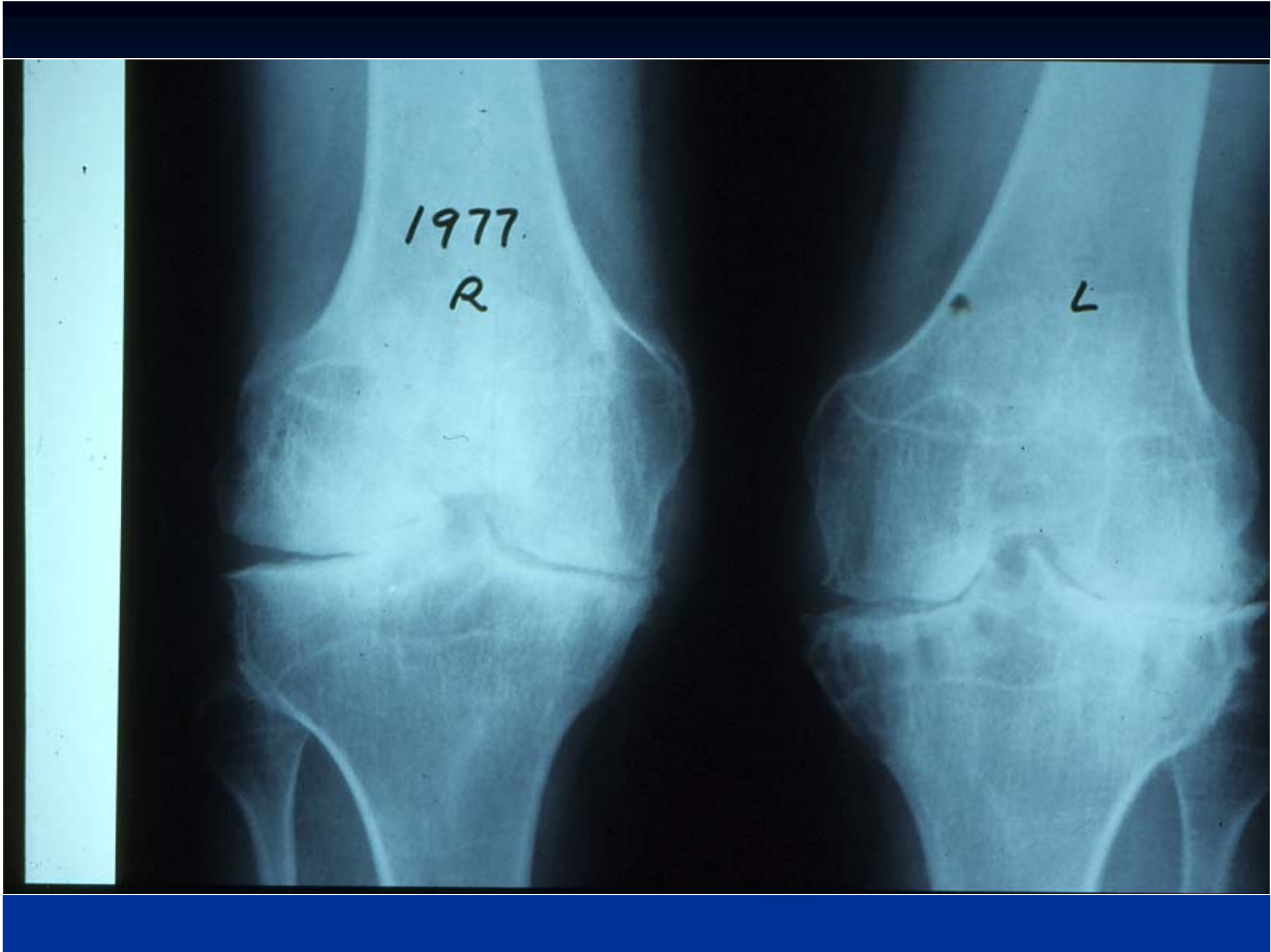
354224R

ANTERIOR VIEW

CHRONIC HAEMARTHROSIS

- SUB ACUTE CHANGES FOR 6/12
- PROGRESSIVE CHANGES UNTIL
END STAGE FIBROTIC
CONTRACTED
TOTALLY DESTROYED
- BLEEDING OFTEN ABSENT OR INFREQUENT IN THE
LATE STAGES.







MANAGEMENT

- PROPHYLAXIS
- ACUTE HAEMARTHROSES
- SUB-ACUTE ARTHROPATHY
- CHRONIC ARTHROPATHY

PROPHYLATIC CLOTTING FACTOR REPLACEMENT

- THREE OR MORE TIMES WEEKLY-
FACTOR LEVEL \approx 10%
- PHYSIOTHERAPY

STRONG MUSCLES

CYCLING

SWIMMING

WALKING

NON-BODY CONTACT SPORTS

ACUTE BLEEDS

1. ADEQUATE HAEMATOLOGICAL CONTROL
2. REST 48 HOURS
3. ASPIRATION
4. MOBILISATION - ACTIVE MOVEMENTS ONLY

ICE PACKS

ULTRASOUND

HYDROTHERAPY

DRUG TREATMENT

- ANALGESICS
- ANTI-INFLAMMATORY AGENTS
NSAID'S, LOCAL I/A AGENTS,
CORTICOSTEROIDS
- DMARD'S, SPECIFIC AGENTS
- IMMUNOSUPPRESSANTS AND CYTOTOXIC DRUGS
- GENERAL SUPPORTIVE AGENTS
 - ANTI-DEPRESSANTS
 - MUSCLE RELAXANTS
 - BONE PROTECTIVE MEDICATIONS

M_x SUBACUTE ARTHROPATHY

Medical synovectomy - synoviorthesis

(DXRT)

radio-colloids (⁹⁰Yt)

(thiotepa)

(osmic acid)

rifampicin

tetracyclines

ethamoline

SURGERY

- TERTIARY HAEMATOLOGY CENTRE
- ADEQUATE SUPPLIES AND HAEMATOLOGICAL CONTROL
- ORTHOPAEDIC SURGEON EXPERIENCED IN HAEMOPHILIA SURGERY



PSEUDOTUMOURS

MASSIVE HAEMORRHAGE

SKIN ULCERATION

GANGRENE

PAIN

SEPSIS

FRACTURE

AMPUTATION

(BOWEL OBSTRUCTION)

PROBLEMS

- INHIBITORS
- SEPSIS
- HIV / HEP C / CJD / ? OTHERS
- THE ANKLE
- THE ELBOW
- OSTEOPOROSIS
- THE ELUSIVE GENE THERAPY

PROBLEMS

- INHIBITORS
- SEPSIS
- HIV / HEP C / CJD / ? OTHERS
- THE ANKLE
- THE ELBOW
- OSTEOPOROSIS
- THE ELUSIVE GENE THERAPY

INHIBITORS

PREVENTION

rh FACTOR VIIa

MASSIVE REPLACEMENT

IMMUNOSUPPRESSIVE AND CYTOTOXIC DRUGS

- RETUXIMAB “MabThera”

PLASMAPHERESIS

PROBLEMS

- INHIBITORS
- SEPSIS
- HIV / HEP C / CJD / ? OTHERS
- THE ANKLE
- THE ELBOW
- OSTEOPOROSIS
- THE ELUSIVE GENE THERAPY

PROBLEMS

- INHIBITORS
- SEPSIS
- HIV / HEP C / CJD / ? OTHERS
- THE ANKLE
- THE ELBOW
- OSTEOPOROSIS
- THE ELUSIVE GENE THERAPY

PROBLEMS

- INHIBITORS
- SEPSIS
- HIV / HEP C / CJD / ? OTHERS
- THE ANKLE
- THE ELBOW
- OSTEOPOROSIS
- THE ELUSIVE GENE THERAPY







PROBLEMS

- INHIBITORS
- SEPSIS
- HIV / HEP C / CJD / ? OTHERS
- THE ANKLE
- **THE ELBOW**
- OSTEOPOROSIS
- THE ELUSIVE GENE THERAPY





PROBLEMS

- INHIBITORS
- SEPSIS
- HIV / HEP C / CJD / ? OTHERS
- THE ANKLE
- THE ELBOW
- **OSTEOPOROSIS**
- THE ELUSIVE GENE THERAPY

PROBLEMS

- INHIBITORS
- SEPSIS
- HIV / HEP C / CJD / ? OTHERS
- THE ANKLE
- THE ELBOW
- OSTEOPOROSIS
- THE ELUSIVE GENE THERAPY

PAIN RELIEF AND DRUG ADDICTION

- NON-ADDICTIVE ANALGESICS
- TRANSCUTANEOUS N STIMULATION
- RELAXATION THERAPY, HYPNOSIS
- PAIN PROGRAMS
- LIAISON PSYCHIATRY CBT
- CONSULTATION WITH LAW
ENFORCEMENT AGENCIES

AN OLD BUT NEW PROBLEM AGEING

- GENERAL MEDICAL PROBLEMS
CARDIOVASCULAR, NEUROLOGICAL
- DEGENERATIVE JOINT DISEASE
HIP, KNEE, SPINE
- TRAUMA, FRACTURES

AN OLD BUT NEW PROBLEM AGEING

- GENERAL MEDICAL PROBLEMS
CARDIOVASCULAR, NEUROLOGICAL
- DEGENERATIVE JOINT DISEASE
HIP, KNEE, SPINE
- TRAUMA, OSTEOPOROSIS, FRACTURES,

AN OLD BUT NEW PROBLEM

AGEING

- GENERAL MEDICAL PROBLEMS
CARDIOVASCULAR, NEUROLOGICAL
- DEGENERATIVE JOINT DISEASE
HIP, KNEE, SPINE
- TRAUMA, OSTEOPOROSIS, FRACTURES,

AN OLD BUT NEW PROBLEM AGEING

- GENERAL MEDICAL PROBLEMS
CARDIOVASCULAR, NEUROLOGICAL
- DEGENERATIVE JOINT DISEASE
HIP, KNEE, SPINE
- TRAUMA, OSTEOPOROSIS, FRACTURES,

ACKNOWLEDGEMENTS

- JANE BLEASEL
- SCOTT DUNKLEY
- STEVE MATTHEWS
- LEONE MUDGE
- KEVIN RICKARD
- (FIONA RENNISON)
- AND...