

Contents

1 Introduction to Hypermobility	1
1.1 Historical Background.....	1
1.2 Rheumatological Manifestations.....	2
1.3 Extra-Articular Manifestations of Hypermobility.....	3
1.4 Late Effects of Hypermobility.....	3
1.5 Measurement of Joint Hypermobility.....	4
1.6 Syndromic Associations of Joint Hypermobility.....	5
1.7 Nosology of the Hypermobility Syndromes.....	5
References.....	8
2 Assessment of Hypermobility	11
2.1 Simple Scoring Systems for Hypermobility.....	12
2.2 The Brighton Criteria for Hypermobility Syndrome.....	14
2.3 General Principles of More Precise Measurement at Selected Joints.....	15
2.4 Back and Spinal Mobility.....	17
2.5 Rotation in the Limbs.....	18
2.6 Movement at the Metacarpophalangeal Joint.....	18
2.7 Joint Proprioception.....	19
2.8 Correlations Between Scoring Systems Used in Assessing Joint Laxity.....	21
2.9 Variation of Joint Laxity Within Populations.....	21
2.10 Clinical Applications of Scoring Systems.....	22
2.11 Joint Hypolaxity.....	23
References.....	23
3 The Molecular Basis of Joint Hypermobility	27
3.1 Introduction.....	27
3.2 The Family of Fibril-Forming Collagens.....	28
3.3 Genes Encoding Type I and V Collagens.....	30
3.4 Biosynthesis of Type I and V Collagens.....	31

3.5	Collagen Fibril Assembly	32
3.6	EDS and Type I Collagen	34
3.7	EDS and ADAMTS2	36
3.8	EDS and Lysyl Hydroxylase.....	38
3.9	EDS and Type V Collagen.....	39
3.10	EDS and Tenascin-X.....	40
3.11	EDS and Type VI Collagen	41
3.12	Relationship Between Elastic Fibre Abnormalities, Marfan Syndrome and EDS.....	42
3.13	Conclusions.....	42
	References	43
4	Biomechanics of Hypermobility: Selected Aspects.....	49
4.1	Mechanical Factors in Joint Mobility	50
4.1.1	Relative Contributions of Different Factors.....	50
4.1.2	Bony Surfaces	50
4.1.3	Collagen.....	51
4.1.4	Neuromuscular Control	53
4.1.5	Proprioception.....	54
4.2	Podiatric Aspects	55
4.3	Lubrication and Stiffness	55
4.3.1	Lubrication of the Synovial Membrane.....	55
4.3.2	Measurement of Stiffness	56
4.3.3	Artificial Lubricants.....	57
4.4	Hypermobility and Osteoarthritis	57
4.5	Prospects for Surgical Intervention.....	60
	References	61
5	Musculoskeletal Features of Hypermobility and Their Management.....	65
5.1	Hypermobility and Hypermobility Syndrome	65
5.1.1	Impaired Healing	66
5.2	Epidemiology of JHS.....	66
5.3	The Clinical Significance of Hypermobility.....	66
5.4	Musculoskeletal Features.....	67
5.5	Hypermobility Syndrome in Children	67
5.5.1	Epidemiology.....	68
5.5.2	Clinical Presentation in Childhood.....	68
5.6	Hypermobility in Adults	73
5.6.1	Prevalence	73
5.6.2	Role of Lax Ligaments	74
5.6.3	Clinical Manifestations.....	74
5.6.4	Articular Features	75
5.6.5	Soft Tissue Lesions.....	76

5.6.6	Chondromalacia Patellae	76
5.6.7	Acute Articular and Peri-articular Traumatic Lesions.....	76
5.6.8	Chronic Polyarthritis or Monoarticular Arthritis in Adults	77
5.6.9	Dislocation of Joints	77
5.6.10	Temporomandibular Joint Dysfunction	77
5.6.11	Premature Osteoarthritis (Other Than TMJ).....	78
5.6.12	Spinal Complications.....	78
5.6.13	Bone Fragility	79
5.6.14	The Natural History of JHS and the Development of Chronic Pain	80
5.7	Management of Articular Complications in the Hypermobility Syndrome	81
5.7.1	General Management.....	82
5.7.2	Specific Management.....	82
5.7.3	Rest	82
5.7.4	Local Steroid Injections	83
5.7.5	Physiotherapy.....	83
5.7.6	General Principles.....	84
5.7.7	Passive Mobilisation	85
5.7.8	Exercise Therapy	85
5.7.9	Podiatry.....	87
5.7.10	Surgical Intervention.....	87
5.7.11	Soft Tissue Lesions	88
5.7.12	Persistent Synovitis.....	88
5.7.13	Recurrent Dislocation or Joint Instability.....	89
5.7.14	Cervical or Lumbar Discectomy.....	90
5.7.15	Surgery of the Foot	91
5.7.16	Advanced Osteoarthritis	91
5.7.17	Symptomatic Treatment.....	91
5.7.18	Analgesic and Non-steroidal Anti-inflammatory Drugs.....	92
5.7.19	Massage, Mobilisation, Hydrotherapy and Water Immersion.....	92
5.7.20	Behavioural Techniques.....	93
5.7.21	Acupuncture and Transcutaneous Neural Electrical Stimulation	93
5.7.22	Denervation Procedures.....	94
5.7.23	Support and Information.....	94
	References	95
6	Extra-articular Manifestations of Hypermobility	101
6.1	Introduction.....	101
6.1.1	Weakness of Supporting Structures Including Pelvic Floor Insufficiency	102
6.1.2	Mitral Valve Prolapse.....	104

6.1.3	Chronic Pain	105
6.1.4	Proprioceptive Impairment	106
6.1.5	Lack of Efficacy of Local Anaesthetics	106
6.1.6	Autonomic Dysfunction.....	106
6.1.7	Certain Psychiatric Disorders	107
6.1.8	Functional Disorders of the Gastrointestinal Tract.....	108
6.2	Straws in the Wind.....	109
	References	110
7	Illustrative Case Histories.....	113
	Case 7.1: A 6-Year-Old Boy with EDS II/III with Grossly Unstable Hind Feet.....	113
	Case 7.2: Pelvic Floor Problems After Childbirth in a Patient with EDS Hypermobility Type.....	115
	Case 7.3: A Labral Tear and Autonomic Dysfunction Complicating Hypermobility	116
	Case 7.4: Complexities in Diagnosis and Management	118
	Case 7.5: Bony Abnormality and Complications of Subluxation	119
	Case 7.6: Arnold–Chiari Malformation and Specialised Physiotherapy	120
	Case 7.7: The Performing Artist.....	121
	References	123
8	Hypermobility in the Performing Arts and Sport	125
8.1	Dancers	125
	8.1.1 Are Ballet Dancers Born or Made	125
	8.1.2 Is Generalised Joint Laxity an Asset or a Liability in Ballet Dancing?	126
	8.1.3 The Prevention of Injury.....	130
8.2	Contortionists.....	131
	8.2.1 Historical Background.....	131
	8.2.2 Nosology and Semantics.....	131
	8.2.3 Training.....	132
	8.2.4 Socio-medical Implications	133
8.3	Musicians.....	133
8.4	Occupational Ills of Instrumentalists	135
	8.4.1 Illustrative Case Histories	137
	8.4.2 Repetitive Strain Syndrome	140
8.5	Sport.....	140
	8.5.1 Joint Hypermobility in Selected Sports	141
	8.5.2 Joint Hypermobility in Cricket	143
	8.5.3 Joint Hypermobility in Yoga.....	143
8.6	Hypermobility and Injury	144
	8.6.1 Training Methods to Improve Joint Flexibility	145
	8.6.2 Hormonal Aspects	146
	References	147

9 Heritable Hypermobility Syndromes	151
9.1 Ehlers–Danlos Syndrome	151
9.1.1 General Features	152
9.1.2 Nosology.....	152
9.1.3 Diagnostic Considerations	155
9.1.4 Rare Forms of the EDS	157
9.1.5 Articular Manifestations	158
9.1.6 Orthopaedic Management of Articular Problems.....	163
9.1.7 Non-articular Complications	164
9.1.8 Resources: Patient Support Groups	167
9.2 Familial Articular Hypermobility Syndromes	167
9.2.1 Nosology.....	168
9.2.2 Articular Complications.....	170
9.2.3 Other Phenotypic Manifestations	171
9.3 Miscellaneous Joint Laxity Syndromes	171
9.3.1 Joint Laxity in Inherited Connective Tissue Disorders.....	171
9.3.2 Skeletal Dysplasias with Predominant Joint Laxity	175
9.3.3 Dwarfing Dysplasias with Variable Joint Laxity	178
9.3.4 Genetic Syndromes in Which Hypermobility Is Overshadowed by Other Manifestations	181
References	183
10 Future Avenues for Research	191
10.1 Arterial Elasticity.....	191
10.2 Cytokine Modulation	193
10.3 Candidate Genes	194
10.4 Disease Association: True or Artefactual?	194
10.5 Neurological Aspects.....	195
10.6 Podiatry.....	196
10.7 Hormonal Aspects	196
10.8 Joint Hypermobility as a Model of Accelerated Osteoarthritis	197
References	197
Index	199