## Frank T Horan

Late Editor and Emeritus Editor of the British Volume of the Journal of Bone and Joint Surgery

Peter Beighton and I met and became friends while students at St. Mary's Hospital Medical School. We played rugby together, participated in car rallies and enjoyed the great social opportunities that were available. After qualification, we played in the Mary's 'B' XV – the veterans' side – and after one particular game, we were sitting next to each other in the bath when Peter, who was a registrar in general medicine at St. Thomas' Hospital, asked what I knew about Ehlers-Danlos syndrome, as he was proposing to try and trace 100 such patients and required help with the skeletal aspects. He approached this task with typical drive and fervour. I saw about 30 of the patients. It became clear to us that some had been misdiagnosed and were simply hypermobile. Some had a family history that appeared to demonstrate autosomal dominance; but a few, often professional contortionists, needed to practise a strenuous stretching regimen in order to maintain their joint laxity. This research project began Peter's interest in clinical genetics. He then spent a year at the Department of Genetics at Johns Hopkins Hospital in Baltimore, under the direction of Victor McKusick. After returning to St. Thomas', he took up a research post in Johannesburg, supervised by Prof. Louis Solomon.

He was then appointed to the Chair in Human Genetics at the University of Cape Town (UCT). Here, he realised that he had access to a huge population base and began to expand his knowledge and expertise. He became particularly interested in inherited bone dysplasias and invited me out on a number of occasions to see patients, review papers, examine radiographs and extend our knowledge of these disorders. We became aware of the work of Sir Thomas Fairbank, whose book, entitled *An Atlas of General Affections of the Skeleton*, was the first attempt to properly classify these rare syndromes. Sir Thomas made meticulous handwritten notes of the patients he had seen, and preserved the radiographs with care. On his death the collection was left to the Department of Radiology at the Royal National Orthopaedic Hospital. We were able to review this work and attempt further classification in the light of more recent knowledge.

Patients with such problems usually present to orthopaedic clinics. However, their rarity is such that an individual surgeon may only encounter one in a lifetime of practice. We therefore felt that it would be useful to produce a short book, outlining and illustrating the features of the more common conditions, based on the records of over 1 000 patients on the Bone Dysplasia Registry in the Department of Human Genetics at UCT. This was published in 1982 entitled *Orthopaedic Problems in Inherited Skeletal Disorders.*<sup>[1]</sup>

When we became too old for rugby, Peter began to increase his running and encouraged me to do so when I visited Cape Town. We subsequently ran several marathons together, but he was always faster and fitter than me at this distance, presumably because of the greater time for training enjoyed by an academic!

In subsequent years, Peter branched out into wider aspects of clinical genetics but has still retained his interest in developmental abnormalities of bone. During my years on the editorial staff of the British Volume of the *Journal of Bone and Joint Surgery* he remained our principal advisor and main reviewer on the genetic aspects of orthopaedics.

1. Horan FT, Beighton P. Orthopaedic Problems in Inherited Skeletal Disorders. Berlin: Springer-Verlag, 1982

## David N Hall

Retired Colonel, Royal Engineers

I first met Peter Beighton in Luluabourg, Congo in 1961 when he was the Medical Officer for a Ghanaian regiment, serving with the United Nations Peace-keeping Force on secondment from the Royal Army Medical Corps. Our paths crossed several times before we were both returned to Ghana and then home to England. Peter clearly had an adventurous spirit and went back to Britain overland across the Sahara, while I spent 3 months leave on camel treks before being posted to Sandhurst as an instructor.

In the months that followed, I relished the breaks from Sandhurst to meet Peter on Saturday evenings in some of the best London pubs near St. Mary's Hospital, Paddington. We made plans for future expeditions with research projects to the Sahara, and the physiological issues of life in this harsh environment were high on the agenda.

In 1962, the British Army was taking acclimatisation to hot, dry climates seriously, so Peter and I decided to investigate this issue when taking a group of young cadets straight from the cool of England into the Libyan desert in the hottest month of the year. Before departure, I had been summoned by the Sandhurst Commandant and told in no uncertain terms that he wanted all the cadets back safely. I relied on Peter, who knew well the dangers of heat illness. All returned safely from Libya and only one cadet had been affected, but was quickly restored in Peter's care.

Typical of Peter, when we were standing not far from the 6 000-ft Gebel 'Uweinat on the border in Sudan, he said to me, 'David, we have the organisation and know-how, why don't we travel on to Ethiopia?' His sense of enquiry and adventure was an inspiration to the whole team.

Then, in 1970, came a major 4-month expedition to the southern Sahara with Peter and a 20-man team. They comprised a nice balance of interactive research disciplines, including archaeologists from Berkeley, USA and an eminent geomorphologist from Australia. Peter provided medical cover and carried out research on psychological factors and the application of a new index of thermal comfort, supported by the late Dr Hugh Bevan-Jones from the University of Birmingham. The Medical Research Council (MRC) funded this research into the stresses inherent in desert exploration.

When the expedition dispersed, Peter set off on a 500-mile journey with a couple of camels and a local Tuareg nomad, in order to measure and compare their respective water intake and output during their 20-day trek. His findings, contrary to general thinking, were that the fundamental metabolic requirements for water were determined by environmental and cultural factors such as the sun cover, activity and diet, rather than genetic influences on kidney function and sweating mechanisms.<sup>[2]</sup>

- 1. Hall DN, Williams MAJ, Clark JD, Warren A, Bradley P, Beighton P. The British expedition to the Air Mountains. Geograph J 1971;137(4):445-467.
- 2. Beighton P. Fluid balance in the Sahara. Nature 1971;233(5317):275-277

## **Steve Mannion**

Consultant Orthopaedic Surgeon, Blackpool Victoria Hospital and Royal Preston Hospital; and Head of the Department of Conflict and Catastrophe Medicine, St George's Hospital, University of London, UK

It must have been the end of the summer term of 1979; an old boy of the school had been invited to give a talk to the prospective sixth formers about his career and work. I was vaguely uninterested - already set on the excitement of a military career, a dull and crusty academic would almost certainly have little to offer.

Blackpool was an already-fading British seaside town of boarding houses and rainy bank holidays; Arnold School, which was the local private school, had recently become co-educational - not your Eton or Harrow, but of solid reputation and respectable exam results.

And so the Prof began, giving not a dull monologue, but an animated and passionate account of his life and work. Science - not the drab science of 1970s school textbooks and musty chemistry labs, but a living, breathing science of real people and real places, combined with clinical medicine; not in white coats and starchy hospital wards, but in the fields and villages of Africa, with expeditions across the Sahara, Easter Island, Tristan da Cunha, as well as 'lobster claw' upper limb deformity, bizarre X-rays of rare skeletal dysplasias, hypermobility syndrome, Huntington's chorea ...

The seed had been sown. My military career was short lived and the transfer to a medical degree challenging but ultimately successful. Basic and higher orthopaedic surgical training combined with missions overseas with the Red Cross and Medécins Sans Frontières; wars in Afghanistan, Ethiopia, Mozambique, Sri Lanka; a terrible genocide in Rwanda; a master's thesis on the biomechanics of landmine injury, then forgoing a straightforward orthopaedic career in the UK to be a missionary surgeon in Malawi for 4 years.

Working as a trauma surgeon in Johannesburg in 1994, one of South Africa (SA)'s most challenging periods, I conspired to travel to Cape Town to meet the source of my inspiration. I was pleased to find Prof. Beighton's passion and zest for life had not been dulled by the intervening years and was struck by the Blackpool and school photographs which adorned his Cape Town office wall.

Peter still visits Blackpool every year, still has good friends there and shares a beer (or two) with me in the local pub. I'm flattered that he invites that schoolboy from the 1970s to do what I can to keep his professorial knees going, ravaged as they have been by using his knees over years of sport and adventure, including competitive orienteering to a high level.

I am eternally grateful to Peter, not only for the inspiration he gave me to pursue a career in medicine, but also the encouragement and faith to pursue a career path 'less travelled', and am honoured to be able to contribute to his Festschrift publication.

## **Donald Basel**

Associate Professor/Interim Chief, Division of Genetics, Department of Pediatrics, Medical College of Wisconsin, Milwaukee, Wisconsin, USA

Peter Beighton has heralded the eponymity of others through his authorship of The Man Behind The Syndrome and The Person Behind The Syndrome. It is thus fitting that he is eponymous for the Beighton Score, a simple yet highly effective scale used to assess generalised joint hypermobility.

In 1973, when the Beighton Score was first described, there would have been no context to comprehend that a Google search would yield in excess of 64 000 hits and that the Beighton Score can be downloaded as a mobile device application to allow self-evaluation. The Beighton Score is the most commonly used and universally accepted scale to assess joint mobility within all subspecialties of medical practice. Hypermobility syndrome, a disorder in which there is significant diagnostic variability, will be the focus for the 2016 Ehlers-Danlos Syndrome International Symposium held in New York City in May 2016.

The prevalence of generalised articular hypermobility is not well documented, with reports varying from approximately 10% to near 40% for adolescents in the general population. The growing body of literature associating hypermobility to a vast array of clinical findings ranging from fatigue, chronic pain, dysautonomia (temperature dysregulation, heart rate variability, functional gastrointestinal disease and postural orthostatic tachycardia syndrome), generalised anxiety disorder and chronic disruptive sleep disorder, speaks to a much greater function for connective tissues in overall wellbeing. Despite significant amounts of study, the exact pathoaetiology of these disparate symptoms remains poorly understood. Many of these findings are common to all hereditary connective tissue disorders, which in turn have distinct phenotypic characteristics enabling classification. Hypermobility is a documented primary feature in 75% of all known connective tissue disorders.

The simple yet reproducible Beighton Score for determining hypermobility will remain a foundation for ongoing study into the ubiquitous impact of connective tissue dysfunction which affects a large proportion of the general population. This is a ripple in the wave of contributions from a prolific academic.