The Man HIMSELF

The scoliosis of King Richard III

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Did he experience pain and disability and what treatment could twenty-first century medicine provide? The discovery of the skeletal remains of Richard III revealed a severe scoliosis, somewhat surprisingly considering his martial prowess. This paper discusses Richard's disability, and contrasts the available management of scoliosis at three points in history: the time of Richard III, the time of Dr Saxton Barton,[1] the founder of The Richard III Society, and the current period.

Introduction

The skeleton discovered under the car park at the site of the previous Grey Friars' church in 2012 was confirmed to be that of King Richard III. The spine had a severe scoliosis, for which there are many therapeutic options today, including the surgery performed on a current member of the royal family. Did Richard suffer pain or disability because of this abnormality?

Before the Plantagenet monarchs

The ancient Hindu epics first described spinal deformities and the use of axial traction as therapy five millennia ago. Hippocrates (460–370 BC) used the term scoliosis, meaning bending, to describe any spinal deformity, including those caused by injury. Galen (130–200 AD) used the term as we do today, to describe lateral curvatures only. The ancient Greeks utilised traction, either on a rack, or by suspending the body vertically, either feet-down or head-down, utilising the body's weight, or spinal compression while lying on a hard board as therapies. Diets, massage and herbal therapy were also applied with little progress over the next 1500 years.[2, 3]

Fifteenth century – the man himself

Although no physical abnormalities were recorded during Richard's lifetime, nor when his naked corpse was displayed after he was betrayed and killed at the battle of Bosworth, in the years after his death there were allegations of skeletal abnormalities. Rous stated that his right shoulder was higher than the left; Thomas More stated that the left was higher than the right, and Polydore Vergil stated that one was higher than the other. Sceptics attributed such ambiguous but malicious statements to Henry Tudor, who was far from being the heir to the throne and had a need to denigrate his predecessor, as we see in politicians today justifying their limited claims to leadership. However, Richard's skeleton revealed a severe scoliosis[4] (Figure 1) which probably developed during adolescence and may well have been visible to those close to him. Scoliosis is detected in children today by simply observing the spine from behind when the child bends over, as shown in Figure 2. It is hard to believe that those who dressed the monarch could not have seen such an abnormality, but no record exists of any abnormality being detected by his adherents during his lifetime.

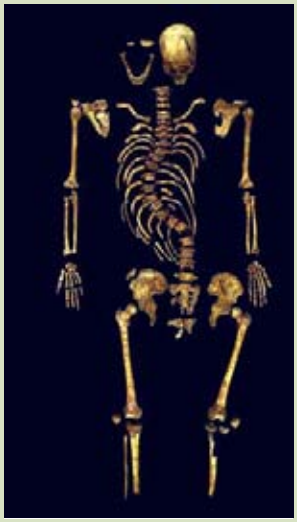


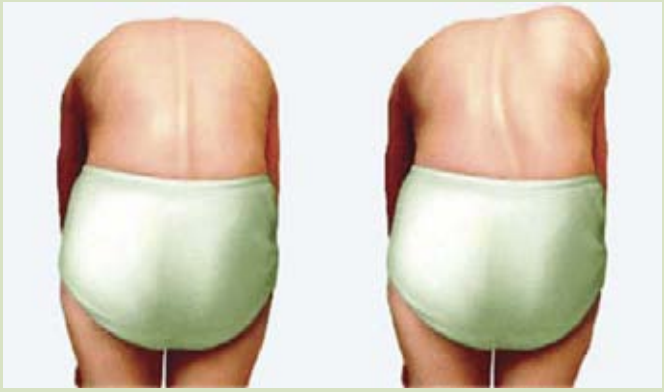
Figure 1. Richard's skeleton.

Diagnosis, definition and measurement of severity

Scoliosis is defined as a 10° or greater lateral curvature of the spine on a coronal radiographic image while standing erect. A simple spinal X-ray remains the gold standard of imaging for the evaluation of scoliosis.[5, 6]

The degree of severity or curvature is measured by the Cobb angle, named after the American orthopaedic

Figure 2. The Adam's forward bend test performed by (left) a patient without scoliosis, and (right) a patient with scoliosis showing a rib prominence.



surgeon (1903–1967). The top and bottom vertebrae of the curved section with the most tilt are identified; parallel lines are drawn along the top of the upper vertebra and along the bottom of the lower vertebra to the point at which they intersect. The angle at which they intersect is the Cobb angle (Figure 3). Richard had a Cobb angle of 85°, indicating a severe scoliosis

Scoliosis may be seen associated with a variety of neurological syndromes and other diseases such as muscular dystrophy and neurofibromatosis and is known as syndromic scoliosis. There is no evidence of Richard having any associated disease. Congenital scoliosis can be seen in neo-natal infants; however, the cause of scoliosis is unknown in most cases, though there is a mild genetic association. Scoliosis is more concordant in monozygotic (identical) twins than dizygotic (non-identical) twins, and certain genetic protein mutations are imputed as possible causes. However, research is ongoing and the known or believed remains of Richard's father, the duke of York, and of his brothers, Edward, Edmund and George, at burial, reinterment or coffin-opening, have no documentation of scoliosis. If not congenital it is then known as idiopathic scoliosis, which is subclassified as infantile between birth and the age of 3 years, juvenile between 3 and 10 years of age, and adolescent, the more common form that is suspected in Richard, after the age of 10 years. This is found in some 2% of adolescents, but only a fifth of these require any treatment.

How does adolescent idiopathic scoliosis present itself?

Most people with scoliosis are identified by an observer, perhaps a relative, doctor or school nurse (or in Richard's case a dresser) or the individual themselves, noting some skeletal abnormality such as uneven shoulders, or prominence of one hip or some ribs (see Figure 2) Few present with pain. The incidence of backache is slightly more than in the general population but is never disabling.

The natural history of scoliosis and the risk factors for curve progression.

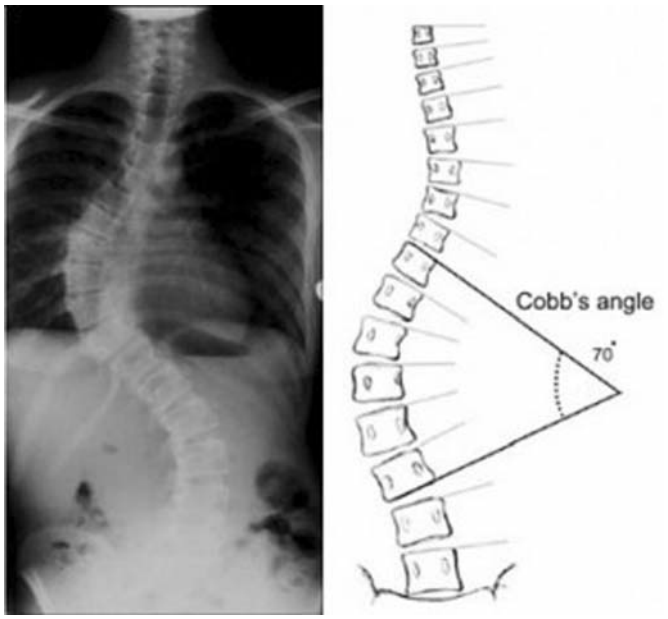
The two- or three-year adolescent growth spurt strikingly increases progression of scoliosis curves, then by the time growth is completed progress slows or ceases. There is a tendency for minor scoliosis, 10–20°, not to progress after the second decade of life, but more severe scoliosis, 40–50°, progresses throughout adult life. Bone maturity and severity of curvature are therefore the major factors when considering spinal surgery. In one series of 205 cases, 16% of those with a curvature of 20° at the onset of puberty, eventually required surgery, but all those with a curvature greater than 30° at the same stage required surgery.[4,5] In another series of 133 patients followed for 40 years, 68% showed deteriorating scoliosis after skeletal maturity. However those with a scoliosis less than 30° did not get worse, while those with curvatures worse than 50° progressed at 1% per year. Infantile and syndromic scoliosis are associated with an increased risk of heart and lung disease, but there is no clear evidence that this is also true of mild idiopathic adolescent scoliosis.

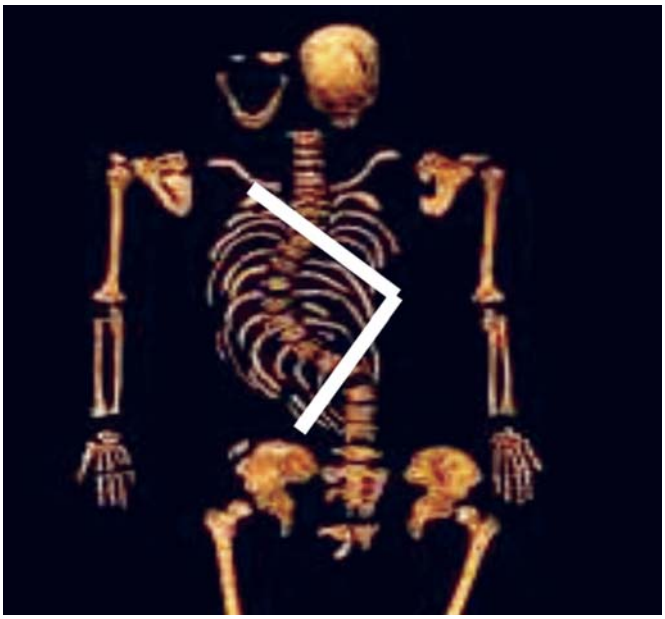
Pain and disability

Speculation has arisen about any symptoms Richard may have experienced, particularly pain, reduced life expectancy or respiratory difficulties. Twentieth-century research answers some of those questions.

A study of 2,442 individuals with scoliosis found that only 23% had pain when scoliosis was first detected, and 9% developed pain later, leaving 68% with no pain.[5,6,7] A third of those with pain also had additional spinal problems, including osteoarthritis, posterior

Figure 3. Cobb angle and Richard's skeleton. © Dr Brett Diaz





vertebral displacement or a spinal tumour, which Richard did not. Many have suggested that Richard would have been in constant severe pain, but this may not be correct. Pain is not a common feature of scoliosis alone and not disabling. Pain from scoliosis is more at the end of the day and gets better with rest.

An alternative perspective on the possibility of Richard suffering severe pain is obtained from a self-selected group of 101 adult patients analysed by Jackson[8] who actually presented with back pain. In this group, aged between 20 and 63, the average age for developing pain was 28, late in Richard's life. The severity of pain correlated with the severity of the scoliosis: those with a 40° curvature had infrequent mild pain, while those like Richard with a scoliosis over 70° mostly had severe daily disabling pain. Scoliosis involving the lumbar spine caused more pain than thoracic deformity. Richard's scoliosis extended from the forth thoracic vertebra to the second lumbar vertebra. The extent and severity of Richard's curvature suggest serious discomfort would have developed had he lived longer.

The other question about reduced health, respiratory problems and shortened life expectancy can only be answer by studies that observe only adolescent idiopathic scoliosis cases over several decades, with cases of infantile scoliosis and scoliosis due to other diseases eliminated from the analysis. A study from Sweden[9] recruited 130 patients with scoliosis between 1927 and 1937 and reported a follow-up study in 1992 of 115 of them – thus some had been followed for nearly 60 years. An increased mortality was found in post-poliomyelitis, infantile and juvenile scoliosis, but not in the 52 individuals with adolescent scoliosis. However, when the cases were subdivided by severity, there was an increase in the death rate, particularly from respiratory disease for those with severe scoliosis, defined as a curvature greater than 70°. The volume of the lungs and the chest doubles during adolescence, hence normal thoracic spinal growth is necessary. Jackson,[8] in his series of adults with back pain, also found that lung capacity decreased with increasing curvature. A pubertal male with a thoracic scoliosis of greater than 50° will have diminished lung volumes and a fifteenfold increased risk for shortness of breath by the age of 30.

Available therapies

Fifteenth century

Little progress in medicine followed for a millennium and a half to the fifteenth century. Lund[4] suggests that Richard could have been treated with massage, herbal applications, traction and a metal or wooded board to wear, little different from Hippocrates time. No effective therapy was available to Richard.

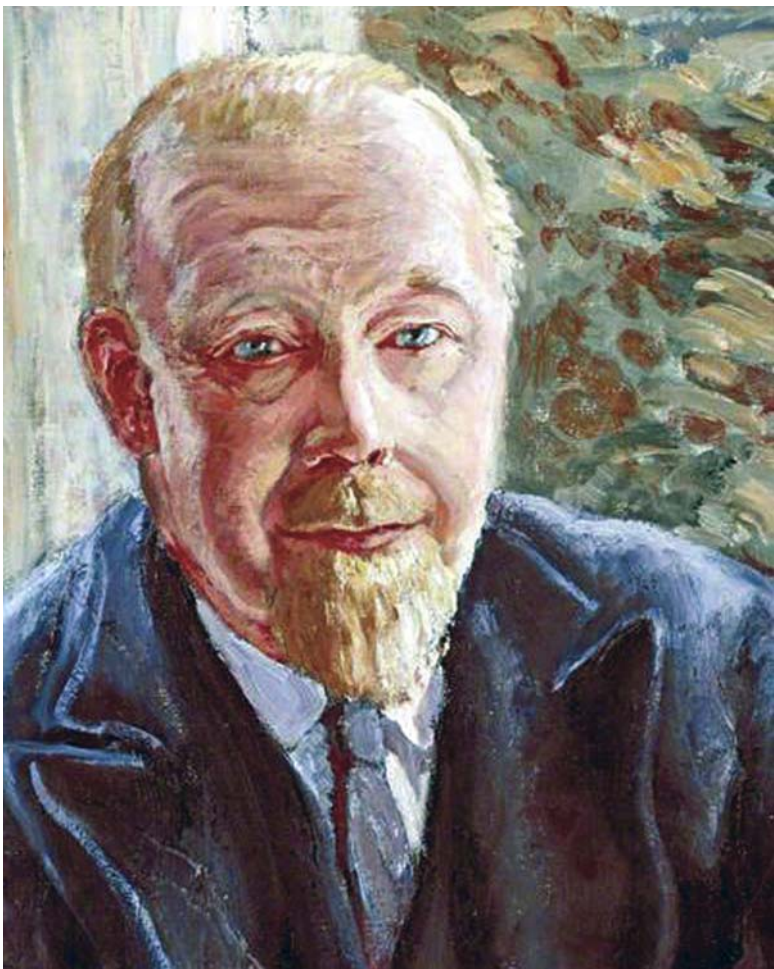
Twentieth century

The Richard III Society was founded in 1924 by the Liverpool consultant obstetrician and gynaecologist, S. Saxon Barton MRCOG (1892–1957), as the Fellowship of the White Boar. Its membership was originally a small group of interested amateur historians whose aim was to bring about a reassessment of the reputation of Richard III. Saxon Barton was a recognised authority on King Richard III and, as secretary of the Fellowship in 1934, erected a stained-glass window in Middleham Church, Yorkshire, to commemorate Richard III. He died following a motor accident in Wales, apparently an unhappy place for Ricardians. Saxton Barton is relevant also because his lifetime encompassed both the initial development of X-rays by Wilhelm Röntgen to the widespread use of radiology in clinical practice, including evaluation of scoliosis, and the description of the Cobb angle to assess severity of scolioses. Robert Lovett was the acknowledged expert in the early twentieth century and recommended forcible correction of scoliosis with the use of plaster of Paris jackets.[10] Early attempts at bone surgery in the early decades of the twentieth century were largely unsuccessful because of recurrence and progress of the curvature.

Twenty-first century

Monday 4 February 2013 was a monumentally important day for Ricardians. The University of Leicester held a press conference to confirm the identity of the Greyfriars car park skeleton as Richard III. Two of the world's

Dr Samuel Saxon Barton (1892–1957), OBE, FRSA, Founder of the Richard III Society. Artist Mavis Blackburn (1923-2005)
Reproduced by kind permission of the Williamson Gallery, Birkenhead.



leading medical journals published articles about idiopathic adult scoliosis very shortly afterwards, the *New England Journal of Medicine* only 24 days later[5] and the *British Medical Journal* on 30 April.[6] Curiously there was no editorial comment on Richard III, but these publications scarcely seem a coincidence. Subsequent clinical information in this article is taken from these two up-to-date and relevant articles.

How is adolescent idiopathic scoliosis managed?

None of the treatments, such as traction or compression, available during Richard's lifetime have any proven benefit in the treatment of scoliosis, and the same is true of many more modern therapies, whether evidence-based such as physiotherapy and dietetics, or the many therapies 'du jour' that lack scientific data.

Casts and bracing

Medical convention recommends plaster casts only for infantile scoliosis. They may reduce curvature progression and delay the need for surgery. It is mostly indicated for flexible curve of 20–30° in a growing child with documented progression of 5° or more, and also some cosmetically acceptable curves with of up to 45°. Surgery is indicated in patients with curves of more than 50°. In pre-pubertal individuals bracing may be beneficial for those with a moderate scoliosis of less than 35°. A brace is usually more beneficial if worn for 23 hours, but part-time protocols are instituted due to compliance issues. In today's world, Richard would have been treated with bracing or surgery during his knightly training, depending on severity of his disease and progression. Richard became involved in the rough politics of the Wars of the Roses at an early age, and became sole commander at age of 17. Given difficult circumstances and the fact that scoliosis is commonly a pain-free condition, he may have ignored it totally.

However, medical knowledge progresses rapidly. A randomised trial comparing bracing with observation for idiopathic scoliosis, published online in the *New England Journal of Medicine* on 19 September 2013, enrolled 242 adolescents with moderate scoliosis defined as a Cobb angle of 20–40°.[11] The brace was a rigid device extending from the chest to the hips. After an average of nearly two years, 52% of the untreated group had progressed to a 50° scoliosis, the point at which surgery is considered as a treatment option, but only 28% of the group wearing a brace deteriorated to this point. The success rate depended on the hours during which the brace was worn. Those wearing the brace for six or fewer hours per day had a 40% success rate, while those wearing it for 12 or more hours a day had a 90% success rate. Richard would have benefited from such a device; perhaps he should have worn his suit of armour for longer periods!

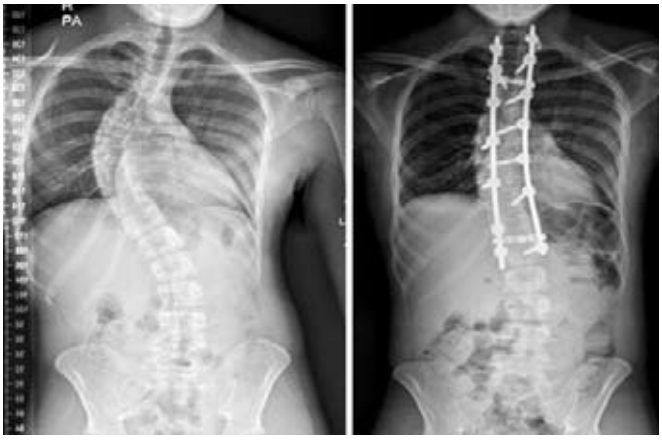


Figure 4. Pre- and post-operative spinal X-rays with correction of the scoliosis

When should surgery be considered?

Surgery is the only proven effective therapy for severe scoliosis in adolescents, originally introduced in the 1960s, involved surgical fusion of the spine with internal fixation with a stainless-steel rod, known as the Harrington rod, to maintain the spine in a straighter position (see Figure 4). Surgery is mostly indicated in patients with Cobb angle more than 50°, increasing curve in growing child or significant cosmetic deformity. In modern era, given the above indications, Richard would definitely have had surgery well before the age at which he died.

Harrington described his new technique as inserting stainless steel rods attached to the outside of the vertebrae which could exert pressure on the spine during surgery to correct the misalignment and maintain that correction.[12] Harrington rods were used for about 30 years, but more modern designs are composed of titanium. Spinal curves of greater than 50° tend to progress slowly after maturity. Therefore a solid surgical spinal fusion is considered for the 10% of adolescents with severe scoliosis, characterised by a Cobb angle curve that is greater than 45–50°, not only to prevent curve progression, but also to improve the cosmetic appearance by correcting the deformity. Unfortunately surgery has not yet been proven to cure back pain, or improve functional status.

Other known cases of scoliosis

Scoliosis appears to be no barrier to physical prowess and reproductive success. Athletes known to have scoliosis include the sprinter Usain Bolt, winner of six Olympic gold medals, and the swimmer Janet Evans, winner of four Olympic gold medals and mother of two children. Elizabeth Taylor, the actress, had eight marriages, seven husbands and four children and was a grandmother by the age of 39. A Princess of York, nineteen generations and over 500 years after Richard, Princess Eugenie, also had a scoliosis for which she had insertion of two titanium rods in 2002 with no reported residual disability.

Conclusion

Current medical opinion indicates that Richard did not have severe back pain, nor any major limitation of activity up to the time of his death. However, he might well have developed increasing respiratory problems during his fourth and fifth decades, with a reduced life span. Today Richard's scoliosis would be regarded as severe, perhaps necessitating a spinal fusion and insertion of titanium rods prior to his involvement in the battles of Barnet and Tewksbury. Perhaps Richard would have preferred a valiant death after a glorious cavalry charge than to suffer a miserable death with increasing pain, disability and breathlessness?

'Richard might well have developed increasing respiratory problems during his fourth and fifth decades, with a reduced life span.'

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A new bust of Richard III

Steve Leadley, a former modeller with Royal Doulton, approached the Society some months ago with a request for permission to be allowed to make a bust of King Richard based upon the facial reconstruction. Following much discussion over the details, the bust has been completed and the Society is offering copies for sale. They will all be hand-made and they will not be mass-produced. Consequently, it will be necessary to take orders and for them to be fulfilled in rotation. Orders will be acknowledged, by e-mail if possible, but members are asked to be patient if they do not receive the bust immediately. They are made in resin with a polished bronze finish and are quite heavy, hence the cost for postage. If you are in a position to wait and collect from a Society meeting, you will save yourself part of the cost. Price, on application, is likely to be around £60.00 plus P&P.

Please make out your cheques to 'The Richard III Society' and write 'R3 bust' on the back.

Orders should be sent to the Sales Officer (address in contacts list on inside back cover)