Rheumatology in art

Benign familial hypermobility syndrome and Trendelenburg sign in a painting “The Three Graces” by Peter Paul Rubens (1577–1640)

Abstract
Clinical features suggestive of hypermobility syndrome and a positive Trendelenburg sign are described in a painting “The Three Graces” (1638–1640) by Peter Paul Rubens, Prado, Madrid. The most obvious findings are scoliosis, positive Trendelenburg sign, and hyperextension of the metacarpal joints, hyperlordosis, and flat feet. The sitters, presumably Hélène Fourment (second wife of Rubens) and her sisters, support the hereditary familial aspect of hypermobility. Manifest hypermobility of the hand has also been found in two other ancient paintings: “Saint Cyriaque” in the Heller Retable by Mathias Grünewald (1450–1528), Frankfurt, and “The wounded man” by Gaspare Traversi, Venice (1732–1769). The finding of signs of hypermobility in ancient paintings shows that artists who are keen observers of nature could describe, or at least record, this condition long before doctors did. The art of the past can be a useful tool in the field of paleopathology.

The hypermobility syndrome, described in 1967 by Kirk, Ansell, and Bywaters,1 as a fruste form of a heritable disorder of connective tissue is now well recognised as a diagnostic entity in rheumatology because of its association with musculoskeletal complaints. The diagnosis of hypermobility is made clinically, without need for costly or invasive testing. Joint hypermobility is defined as abnormally increased mobility of small and larger joints beyond the limits of their physiological movement. The entity is common in young women, with an estimated prevalence of 5% in the healthy adult population.

During my search for paleopathological evidence of rheumatic disorders in ancient paintings, I was struck by the painting of Rubens “The Three Graces”, Prado, Madrid, Spain (1638–1640) (fig 1). One would not expect any physical abnormality in three young sensual graces of Greek mythology, representing Euphrosyne, Aglaia, and Thalia, the three daughters of Zeus, who are the accompanying ladies of Venus, the goddess of love.

The grace in the middle shows clearly a manifest S-shape scoliosis and, interestingly, a positive Trendelenburg sign. She is standing on her left leg and the right buttock is descending instead of lifted upwards as it would be in a normal person.

The test, described by Trendelenburg, a German surgeon (1843–1924), is positive when the pelvis drops on the side opposite to the weight bearing leg. This test is a measure of the patient’s ability to use the abductor muscles of the hip properly. Normally, when a patient bears the weight of the body on one leg (as the girl in the middle of the painting does), the pelvis on the opposite side may be slightly raised by the contraction of the abductor muscles of the weight bearing leg in order to maintain balance. In our subject, who is standing on the left leg, the right hip is descending rather than rising. A positive Trendelenburg test, as seen in this case, is found in patients with a dislocation of the hip, non-union of the femoral neck, coxa valga, arthritis of the hip and sacroiliitis, or marked weakness of the hip abductors.

Further inspection of the painting shows that the grace on the left has fingers in hyperextension at the distal interphalangeal and metacarpophalangeal joints of the fourth and the fifth finger and a flat foot. All the three graces have hyperlordosis of the lumbar spine.

If all these observations were found in one person, the working diagnosis of hypermobility syndrome could explain the findings, but the features appear separately in the three graces. If the graces were sisters or from the same family stock, the scoliosis and the positive Trendelenburg sign could go with the finger deformity seen in the other grace and the hyperlordosis in all of them under the heading of benign familial hypermobility syndrome.

Scoliosis and hyperlordosis are a common feature of benign hypermobility syndrome, and the positive Trendelenburg sign might be due to weakness of the hip girdle muscle. The main symptoms of benign hypermobility syndrome are passive dorsiflexion of the fifth metacarpophalangeal joint to 90°, apposition of the thumb to the
flexor aspect of the forearm, hyperextension of the elbow beyond 90°, hyperextension of the knee beyond 90°, forward trunk flexion placing hands flat on floor with knees extended.

Further research into the history behind the painting confirmed that our working diagnosis was correct. The sisters of this painting of Rubens are, according to Vlieghe,³ Rubens’ second wife Hélène Fourment and her sisters. The same sisters figure in Rubens’ painting “The judgement of Paris”, Prado, Madrid (1638–1639).

The year 1630 was the time of Rubens’ second marriage to Hélène Fourment, who was 16 years old and the youngest of 11 girls. Rubens died in 1640, just after finishing “The Three Graces” and “The judgement of Paris”.

If two of the graces are sisters of the second wife of Rubens, then they are likely to have the same genetic traits. The working diagnosis of familial benign hypermobility is now more appropriate, with the clinical findings of scoliosis, hyperextension of the fingers, flat feet, hyperlordosis, and a positive Trendelenburg sign.

The latter observation—a positive Trendelenburg sign—is not classically described but may fit as a consequence of lax joint instability.

Hypermobility syndrome is a benign genetically determined disorder of connective tissue related to Marfan and Ehlers-Danlos syndrome, characterised by hyperlaxity of the joints, the skin, and in some cases associated with varicose veins, rectal/uterine prolapse, and asymptomatic mitral valve prolapse. Because of joint laxity, the hip can be unstable in the hypermobility syndrome and explain the positive Trendelenburg test. The observed scoliosis is a recognised part of the syndrome because of joint laxity at the spinal level. In hypermobility syndrome, the scoliosis is functional, but this cannot be tested on a static image. If the girl with scoliosis on clinical examination could bend forward then we would see in the case of hypermobility syndrome a correction of the scoliosis with no asymmetry of the thoracic wall, indicating a functional, and not a fixed, scoliosis.

Patients with hypermobility present with a variety of readily identifiable traumatic and overuse lesions, such as traction injuries at tendon or ligament insertions, joint or tendon sheath synovitis, chondromalacia patellae, rotator cuff lesions, or back pain due to soft tissue injury or disc prolapse. Other shows the effects of joint instability, such as flat feet, recurrent dislocation or subluxation—notably, of the shoulder, patella, metacarpophalangeal joints or temporomandibular joints. Others develop a chronic arthritis—either a low grade inflammatory synovitis of traumatic origin (which may mimic or subsequently be misdiagnosed as rheumatoid or juvenile chronic arthritis) or osteoarthritis, which is held by many authorities (albeit on circumstantial evidence) to be a direct complication of hypermobility syndrome.

A link between hypermobility and fibromyalgia has been suggested recently, by triggering disordered pain response in some patients. Patients with hypermobility have, as a group, also some degree of osteopenia.

I have noticed other examples of hypermobile joints of the hands in the paintings of Mathias Grünewald, “Relatable Heller Saint Cyriaque” (1450–1528), Frankfurt, Stadli-ches Kunstinstitut, and “The wounded man” of Gaspare Traversi (1732–1769), Academia di Belle Ari, Venice. Peter Paul Rubens was known (from letters written between 1623 and 1640) to have rheumatic disease. This affected mainly the hand and foot, was chronic in nature, and was complicated by flares of more than one month’s duration, so that he was seriously disabled. The diagnosis is probably gout.⁴ Appelboom et al studied his paintings carefully and found several indications of rheumatic deformities in his paintings, such as the swan neck deformity in the painting of “The Three Graces” and a subluxation of the right wrist in the painting of “Hélène Fourment holding a glove” (1630), Bayerische Stads-gemäldesammlung, Munich, and thought that Rubens projected his rheumatic disease onto his models.⁵ However, the swan neck deformity in the left arm can be interpreted differently. Swan neck deformities may also be an expression of hyperlaxity or hypermobility of the finger joints, as seen in Jaccoud’s syndrome, secondary to inflammation of the ligaments without joint destruction, such as in the older days in acute rheumatic fever and now in SLE. Secondly, swan neck deformities may be present in the hypermobile syndromes, in particular, the benign familial hypermobility syndrome, Marfan and Ehlers-Danlos disease, in which a generalised hereditary collagen defect is the cause of the laxity.

The description of this “case report” of the benign familial hypermobility syndrome in “The Three Graces” of Rubens, presumably Hélène Fourment and her sisters, who lived 400 years ago, supplements a previous report of hypermobility in Taino art (Dominican Republic).⁶ Observation has a key role in clinical medicine, but the paleopathological observation in art shows us how artists, keen observers of nature, could “describe”, or at least record, these conditions long before doctors did.