

# A possible case of dyschondrosteosis in a bog body from the Netherlands

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## Abstract

*In 1951 peat cutters unearthed the bog body of an adult female dating from the Roman period (AD 78-233) in the 'Damsel's Bog' northwest of the villages of Aalden and Zweeloo (province of Drenthe, the Netherlands, fig. 1).*

*The degree of preservation the body has now been assessed using atomic force microscopy imaging (AFM) and histology. AFM images of the skin showed evidence of moderate decomposition of collagen fibrils. Although histology revealed moderate decalcification of the bones, the abdominal organs were found to be very well preserved.*

*Apart from shrinkage and deformation caused by the long immersion in the bog, the Zweeloo Woman's skeleton shows possible signs of a pathological disorder affecting both the forearms and the lower legs.*

*The long bones were measured, the woman's stature was assessed and a CT scan was carried out to assess the degree of micromelia and the possibility of dwarfism.*

*Although shrinkage had caused overall shortening and deformation of single bones (i.e. the pelvis, calcanei and the femora) altering the stature, the radiological findings show probable evidence of Léri-Weill dyschondrosteosis (DCS). DCS is a dominantly inherited dysplasia characterised by short stature with mesomelic shortening of middle segments of the forearms and lower legs. Only three cases of probable or indicative DCS have so far been diagnosed from pre-modern societies. Here we propose evidence of a probable case of DCS syndrome in a bog body dating from the Roman period.*

*It has often been suggested that a substantial number of the individuals that have been found in peat were killed before being deposited in the bog. The Zweeloo Woman's bones show at least 21 cut marks made by a short blade instrument. There is, however, no evidence of any trauma, except possibly in the posterior aspect of the left shoulder, on the outer skin surface. Whether Zweeloo Woman was intentionally killed or died a natural death still remains unclear.*

**Keywords:** Bog bodies, taphonomy, palaeopathology, dyschondrosteosis, The Netherlands.

## 1 Introduction

The term 'bog body' usually refers to human remains found in wetland peat deposits, although other mammals have also occasionally been discovered in such deposits.

Peat is formed at varying rates through compaction and accumulation of plant biomass. Depending on the underlying geology, the available nutrients, topography, the water table level, drainage, and precipitation, raised or blanket bogs may be formed. They may be fairly small or extend over a considerable area. The depth of the peat may vary from 30 cm to over 5 m (Brothwell & Gill-Robinson 2002, 122-123).

The bodies (mummies and skeletons) that have been unearthed at such wetland sites are of great relevance as they provide information on the long-term taphonomy of human remains in these environments. They also provide information on a range of traumata and pathological conditions in a series of individuals showing great variation in preservation (Brothwell & Gill-Robinson 2002, 120; Lynnerup 2007, 2010).

Peat has been cut for fuel for centuries. That makes it likely that human remains would have been discovered in bogs over a long period of time. However, it is only in the past two centuries that literacy and interest in such finds have resulted in a growing number of written records about bog bodies. Based on Dieck's catalogues (1965 and 1986), it was previously stated that the overall number of recorded finds is probably around 2000 (Brothwell & Gill-Robinson 2002, 120; Gill-Robinson 2003, 43). However, critical revision of Dieck's work has shown that most of the bodies in his paper are fabricated (Van der Sanden & Eisenbeiss 2006, 111-122). The real number of discovered bog bodies and bog skeletons is unknown.

The most famous continental bog mummies have been found in parts of northern Europe, especially Britain and Ireland, Denmark, northern Germany and the north of the Netherlands. They range in date approximately from 500 BC to AD 500 (Lynnerup 2010).

The Drents Museum in Assen, province of Drenthe, the Netherlands, has in its collection several bog bodies, among which are the remains of a body found around sixty years ago. These remains consist of a fairly complete skeleton, large soft tissue parts and even viscera. The body is referred to as 'Zweeloo Woman' because it was discovered in what was then the municipality of Zweeloo (now the municipality of Coevorden). Since her discovery she has been the object of several studies. This paper reports current research results obtained by using new techniques, and compares those results with the conclusions of earlier investigations.

### 1.1 Discovery

On December 5 1951, peat cutters working in a bog known locally as *Juffersveen* ('Damsel's Bog') discovered a corpse (Van der Sanden 1990a, 70). The Mayor of Zweeloo was informed of this remarkable discovery and he contacted the *Biologisch-Archaeologisch Instituut* (Institute for Biology and Archaeology)<sup>1</sup> of Groningen University. The following day, professor Dr A.E. van Giffen and his assistant W. van Zeist visited the site. They soon discovered that they were too late to make in-situ observations, the peat cutters having already dug up the human remains. No remains of clothing were observed.

All the archaeologists could do was to collect the bones and soft tissues and study the peat section at the site of the find. The human remains were placed in a large zinc box and taken to Groningen. At the time of discovery, little information was recorded on the findspot of the Zweeloo bog body.

In his daily reports, Van Zeist refers to 'Juffersveen' and 'a small peat bog', but unfortunately without specifying its size and location.

Thanks to the information provided by the then Mayor Greebe, the site of the former bog was located on a map in 1988 (Van der Sanden, 1990a, 70). It lies in an area known as Alder

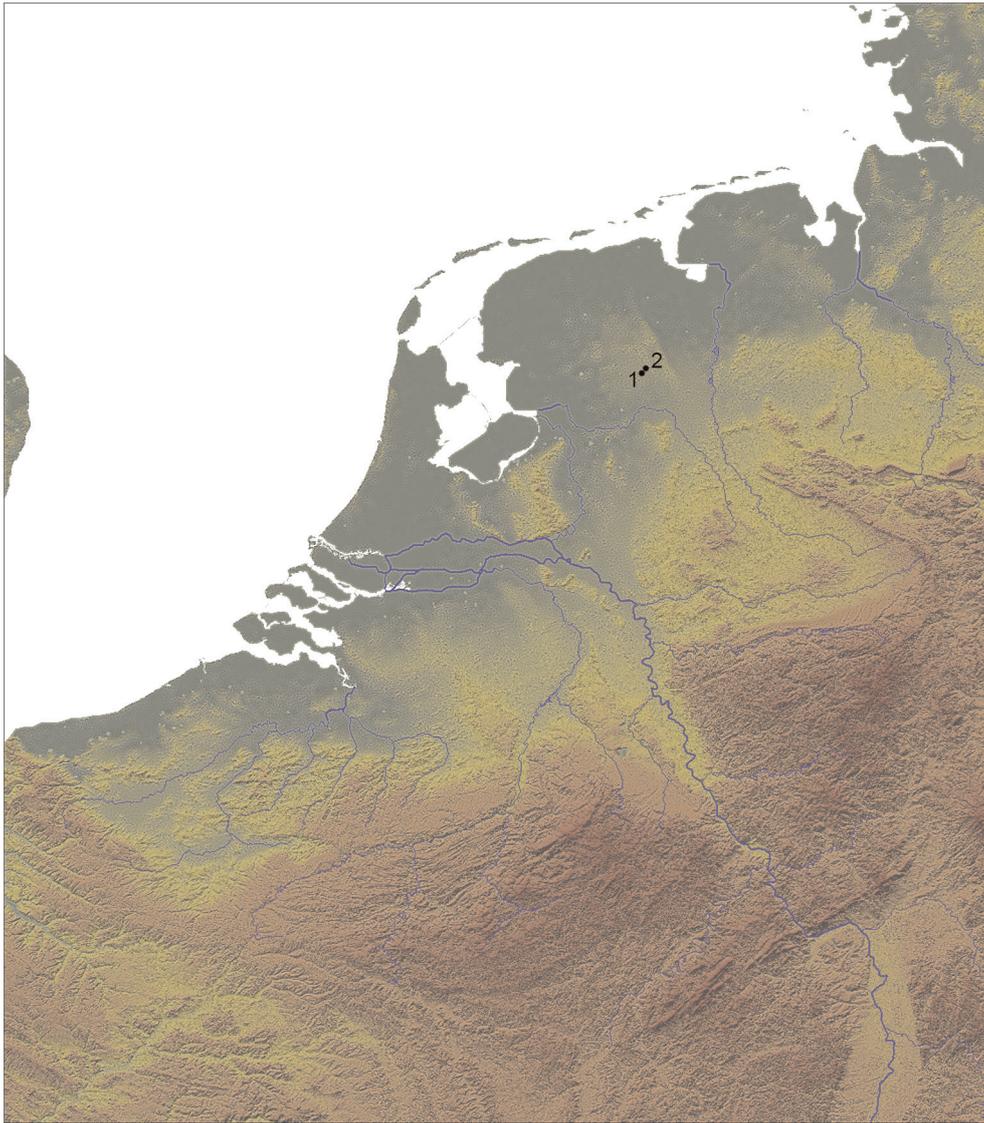


Figure 1 Location of Aalden and Zweelo.

Veld, northwest of the village of Aalden (municipality of Coevorden). Recently, members of the local historical society were asked about the location of 'Juffersveen' and informed one of us (WVDS) that it lay a little further south than the area indicated by Greebe. On a land-consolidation map they sketched a bog measuring around 400 x 200 m extending on either side of Gebbeveenweg.<sup>2</sup>

Their bog coincides reasonably well with an elongated, NW-SE-oriented bog measuring approximately 350 x 125 m which is indicated on a topographic map from around 1900 (Wieberdink, 1900, sheet 224).<sup>3</sup>

If this is indeed 'Juffersveen', and considering that Mayor Greebe indicated the findspot as lying immediately west of Gebbeveenweg, it is likely that the body was buried in the central part of the bog.

The findspot of Zweeloo Woman lies within the boundaries of the village territory (*Marke*) of Aalden, which dates back to at least the Middle Ages, and probably a long time before then. The eastern boundary is formed by the brook valley of the Aelder Stroom. The Roman-period settlement in which the woman may have lived has not yet been found; it is most likely still hidden beneath the plaggen soil of 'Aalder Esch' south of Aalden, where an early medieval cemetery has been excavated. The distance between the bog in which Zweeloo Woman was discovered and her (assumed) settlement is about 2.5-3 km.

## 1.2 Post-excavation studies (1952-1994)

In 1952 Van Zeist published a short article in German on the bog find. He stated that analyses of the human remains at the *Laboratorium voor Anatomie en Embryologie* (Laboratory for Anatomy and Embryology) had shown that they represent a female who was approximately 170 cm tall. Unfortunately the name of the anatomist involved is not reported. Nor are there any written reports giving the exact measurements of the long bone segments.<sup>4</sup>

Van Zeist also discussed the pollen diagram, noting a remarkable gap: Subatlantic peat lying directly on top of Boreal peat. He wrote that he took a pollen sample from 'between the toes of the undamaged foot'. The results of the analysis confirmed the peat cutters' statement that they had found the body in the lower layer of blackish brown peat. According to Van Zeist this implied that the woman had lived in the first centuries AD.

Van Zeist lastly mentioned that the intestines contained many husks of millet (*Panicum miliaceum*) - an observation that did not contradict the find's palynological date. Four years later Van Zeist (1956) published some new information. The woman's hair had been cut around the time of her death and her body had been deposited in an old turf cutting (which would explain the conspicuous hiatus in the pollen diagram). The time of deposition, he estimated, was after AD 200.

### 1.2.1 1987-1993

In the following decades no attention was paid to Zweeloo Woman. In 1987, one of the authors (WVDS), at that time curator of archaeology at the Drents Museum, launched a major bog body research project (Van der Sanden 1990). The first and main aim was a totally new analysis of the preserved corpses - Yde Girl, the Weerdinge Men, Zweeloo Woman and several others - and the finds associated with them (Van der Sanden 1990b, 89-90). Many specialists contributed to this project. We summarise the results below.

The physical anthropologist Dr H.T. Uytterschaut of the Laboratory of Anatomy and Embryology of Groningen University analysed and described the remains (Uytterschaut 1990b, 115-117, 122-123). Due to some confusion resulting from poor documentation by the museum, parts of her conclusions later had to be rectified (explained in Van der Sanden *et al.* 1991-92). On the basis of the shape of the pelvis and the size of the mastoid process, Uytterschaut confirmed the earlier conclusion that the individual was female. Her age at the time of death was estimated on the basis of various observations. Epiphyseal union in the long bones, pelvis and scapula indicated a minimum age of 25, the morphology of the pubic symphysis pointed to an age of between 30 and 60 and the sutures of the skull implied an age of between 20 and 30. Histological analysis (Uytterschaut 1985) of a thin section of the shaft of the left femur indicated that she died at an age of  $34 \pm 7$  years. All this evidence led to the conclusion that the age at the time of death was probably around 35 years (Uytterschaut 1990b, 117).

The marked differences between some of the bones of the left part of the skeleton and their counterparts on the right side (*e.g.* the pelvis) caused a lot of discussion, as the observations were not in accordance with any known physical disorder. Post-mortem deformation (pseudopathology) was suggested as the most likely explanation (Uytterschaut 1990c, 132-135). On the basis of the bones of the right side of the body, the individual was estimated to have been around 138 cm tall, but it was simultaneously assumed that this would not be entirely correct due to post-mortem shrinkage (Uytterschaut 1990a, 102). It was also noted that the woman's hair was cut around the time of her death (Uytterschaut 1990d, 136).

Because of the aforementioned confusion, Dr M. Voortman and his associates of the *Laboratorium voor Gerechtelijke Pathologie* (now *Nederlands Forensisch Instituut* / Dutch Forensic Institute) in Rijswijk re-examined the skin in 1993 and identified the vulva and a breast (right side), which is consistent with the result of the sexing of the skeleton.

The other information can be summarised as follows. Zweeloo Woman was probably of blood group O (Connolly 1990, 148) and suffered parasitic infections caused by whipworm (*Trichuris*) and mawworm (*Ascaris*) (Paap 1990, 164-166).

Her last meal consisted of a gruel whose main ingredient was coarsely ground common millet (*Panicum miliaceum*). The presence of a large quantity of blackberry pips (*Rubus fruticosus*) indicates that she died in late summer/early autumn. In addition, the intestines contained remains of *Polygonum lapathifolium* (curlytop knotweed/pale smartweed), *Polygonum aviculare* (knotgrass), *Brassica* sp. (black mustard or rapeseed), *Linum usitatissimum* (common flax), *Bromus* sp. (grass), *Triticum* sp./*Secale* sp. (wheat or rye), *Hordeum* sp. (barley), *Avena* sp. (oats), a few animal hairs and the wing case of a beetle (*Tenebrio obscurus*) (Van der Sanden 1990d, 136; Hakbijl 1990, 170-171; Holden 1990, 265-269). Analysis of pollen samples from the intestines confirmed the macroscopic results (Troostheide 1990). No DNA was found in the skin or kidney (Osinga & Buys 1990; Osinga *et al.* 1992).

Both the skin and the skeleton were radiocarbon dated. The date of the skin is  $1835 \pm 40$  BP (GrN- 15458), that of the skeleton  $1940 \pm 70$  BP (OxA-1727) (Van der Sanden 1990c, 98). The average of the two aforementioned radiocarbon dates is  $1861 \pm 35$  BP (calibrated  $2\sigma$ : 78-233 cal AD),<sup>5</sup> which means that Zweeloo Woman lived in the Roman period.

### 1.2.2 1994

In 1994, when consulted by one of us (WVDS), Dr R.W. Stoddard of the Department of Pathological Sciences of Manchester University expressed his doubts about the deformed bones being simply a case of pseudopathology. He closely examined the skeleton, measuring and weighing the bones and subjecting some of them to soft X-rays and tomographic scanning, and came to the conclusion that the asymmetry is indeed a post-mortem development, but the short forearms and lower legs are on the contrary to be classified as a pathological phenomenon (fig. 2).

The conclusion of his report (Stoddard 1995), in which he describes the skeleton as that of a woman aged 16-24, is as follows.<sup>6</sup>

'The cadaver shows shortening of the forearms and lower legs by about 50% relative to normal, and corresponding low weights of demineralised bones of these parts. Apart from a mild degree of malformation of the radii, the affected bones are short rather than distorted. There is no remaining evidence of nutritional deficit or infectious illness and no other part of the skeleton appears to be affected. The girdles, proximal portions of the limbs, hands and feet show no abnormality of size, though there is evidence of reactive change in the bones of the feet consequent upon the inevitably abnormal gait.

Several conditions can be excluded. This is not a case of hypopituitary dwarfism, since it is not a generalised failure of growth. Similarly, it is not a phocomelic type of dwarfism, since only a specific segment of each limb is involved and the skull is unaffected. The normal hands and feet argue against an effect of the 'thalidomide-injury' type, for example following a viral infection. An 'anti-Marfan' type of syndrome can also be excluded, since the equivalent segment is involved in each limb and the skull, hands and feet are spared. There is no evidence of nutritional or infectious cause and the localised nature of the abnormality argues against these. The most probable class of disorder in this case is dyschondrosteosis. This develops during later childhood and adolescence, for unknown reasons, producing the type of malformation seen here. The name is purely descriptive and may cover several syndromes. The condition is rare and the origin (or origins) appear to be genetic' (Stoddard 1995, 9).

The diagnosis was quoted in the volume accompanying the bog body exhibition that was held in Silkeborg, Denmark, in 1996 (Van der Sanden 1996, 140-141).

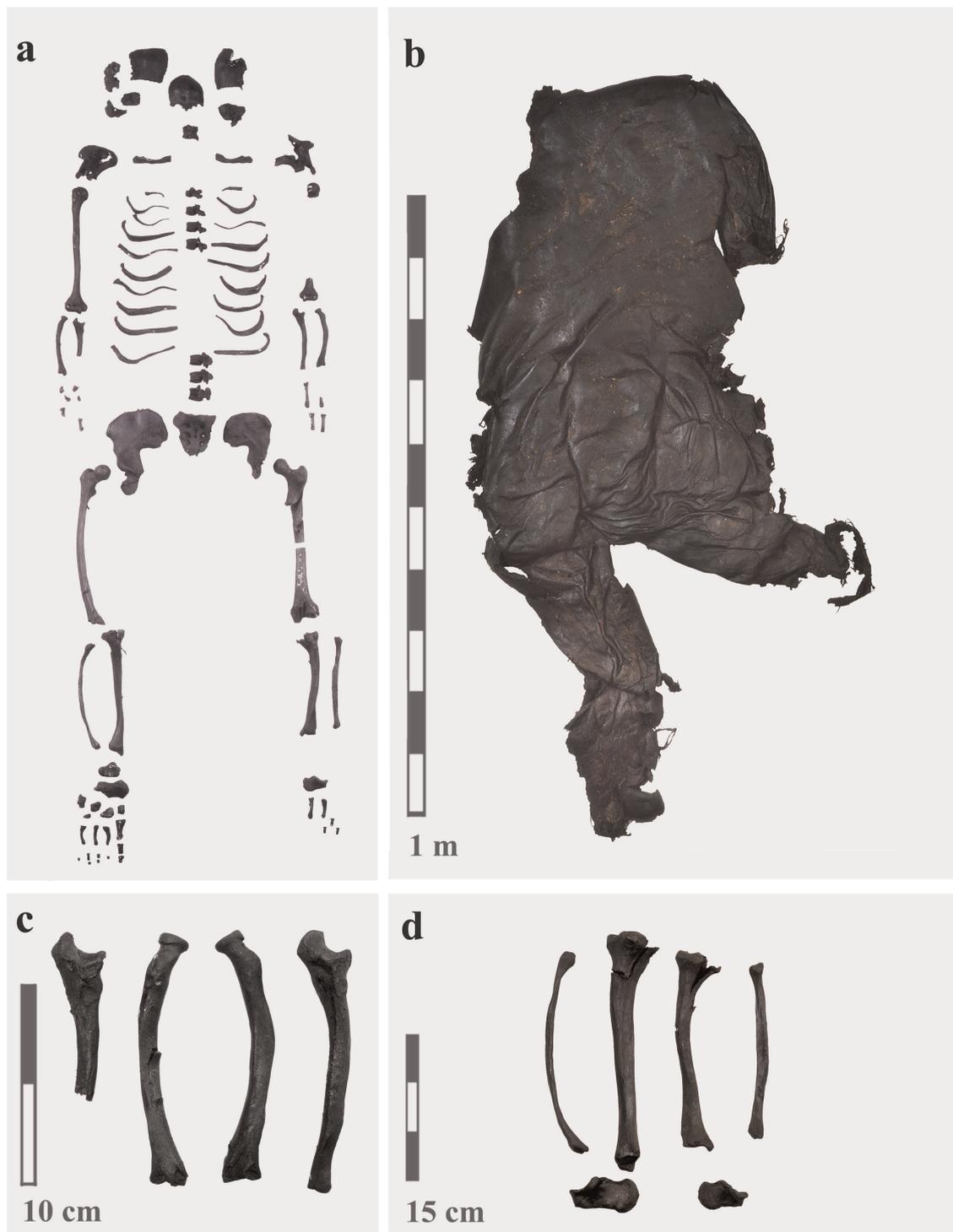


Figure 2 Zweeloo Woman's preserved skeleton (a) and skin (b). (c) and (d) show the shortened, bowed forearms and lower legs.

## 2 New investigations (2009-2011)

Since 2009, a multidisciplinary team of researchers has been re-examining the human remains of Zweeloo Woman to verify the overall state of preservation of both hard and soft tissues and to confirm or reject the previous tentative diagnosis of dyschondrosteosis.

An important question in the interpretation of the human remains is their possible post-depositional deformation and shrinkage.

The preservation of the skeleton along with muscle and ligaments is surprisingly variable in bog bodies. The bones and teeth of most of the skeletons from bogs are severely decalcified because of the acidic environment (Brothwell & Gill-Robinson 2002, 123). Demineralisation is due to the diagenetic influence of the bog acids, which leach minerals (calcium) from the bone. This leaching involves all bony structures, but may affect them to different degrees (Lynnerup *et al.* 2007, 114; Villa *et al.* 2011, 167).

Bones of bog bodies are often distorted and compressed during interment, as observed in the cases of Lindow II and Lindow III and Grauballe Man (Pyatt *et al.* 1991; Schilling *et al.* 2008; Lynnerup *et al.* 2007, 114-120). In the case of Huldremose Woman, anomalous curvature of the bones of the forearm and femur was caused *post mortem* by decalcification and pressure of wood within the peat (Brothwell & Gill-Robinson 2002, 128; Jurik 2007, 99).

Many bog bodies also prove to have shrunk. Because most of these bodies were allowed to dry naturally, they show considerable shrinkage and deformation in single bones or even entire limbs (see Van der Sanden 1996, 18, fig. 15, Lynnerup *et al.* 2007, 119; Lynnerup 2010, 444-445).

Histological analysis was performed to assess the state of preservation of bone and viscera in Zweeloo Woman (section 3.1). The level of skin tissue preservation was assessed by means of atomic force microscopy (AFM) (section 3.2). The results were then used in a metrical analysis to arrive at a more precise estimate of stature and new estimates of body proportions. Lastly, a preliminary review of the observed abnormalities, especially of the forearms and lower limbs, is given (sections 3.3 and 3.4). CT scanning led to a more precise identification and specification of the previously described abnormalities, additionally revealing a series of cut marks on the bones.

## 2.1 Methods

### 2.1.1 Histological analysis

A bone sample was analysed according to the protocol outlined by Maat *et al.* (2000); the intestine samples, liver and kidney, were typed according to their shape and internal position in the mummy's bundle of viscera (Van der Sanden 1996, 116). Small tissue biopsies (0.5 cm x 0.5 cm) that were macroscopically identified as liver and kidney tissues were analysed according to the methods described in Mekota & Vermehren (2005).

After rehydration in Solution III for 48 hours, samples were fixed for 24 hours in 4% formaldehyde, dehydrated and finally embedded in paraffin blocks. The embedded samples were cut into 3µm-thick sections using a microtome (Leica, RM2245). The paraffin sections were histochemically counterstained with either haematoxylin and eosin stain (H&E) or Gram stain (Mullisch & Welsch 2010, 214; 243-244).

### 2.1.2 Atomic force microscopy

Atomic force microscope (AFM) imaging, a well-established technique for obtaining information on the surface properties of a sample, was used to determine the collagen preservation in histological skin samples of the mummy.

2-4µm-thick transverse sections of Zweeloo Woman's skin were processed as described in section 2.2.1, applied to glass slides, dewaxed and rehydrated in a descending alcohol series. The samples were then analysed using an atomic force microscope (NanoWizard®II, JPK Instruments, Berlin, Germany) operating in intermittent contact mode. Measurements were performed in ambient conditions. Silicon cantilever (BS Tap 300, Budget Sensors, Redding, USA) with typical spring constants of 40 N/m and nominal resonance frequencies of 300 kHz were

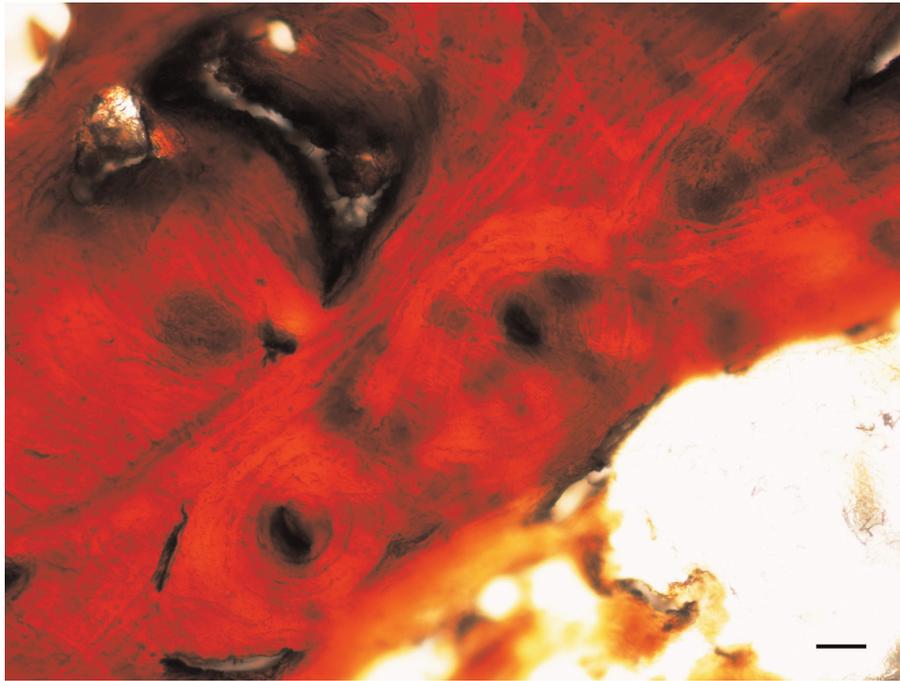


Figure 3 Ground section of the bone tissue. Bar = 20µm.

used. The tip radius was smaller than 10 nm. Image analysis was carried out using SPIP (SPIP 5.0.1.0, Image Metrology, Denmark).

### 2.1.3 CT scanning

A single radiographic evaluation of the entire skeleton was performed in 16 layers using a CT scanner (CT Gemini TF, Philips, NL). The scanning parameters were: 120 KV; 98 mAs; slice thicknesses 0.8 and 1 mm; reconstruction interval 0.75 mm, rotation time 0.5 sec; filter D, image matrix 512x512. The total number of axial slices was 1378. Post-processing, including axial scans evaluation and multiplanar reconstructions, was performed using a Workstation EBW Brilliance (Philips, NL).

## 3 Results

### 3.1 Histological analysis of bones and viscera

#### 3.1.1 Bone preservation

In spite of demineralisation and various damaged areas, histology performed on Zweeloo Woman's bone fragments showed that important cellular structures such as Haversian canals and the lamellate structure have survived (fig. 3) (Pyatt *et al.* 1991).

There is no evidence of generalised destruction caused by mineral dissolution. Such alteration is characterised by the general loss of identifiable features such as bone lamellae, osteocytes and canaliculi (Hollund *et al.* 2011).

The high degree of preservation of the bone's microstructure may indicate that the Zweeloo body was recovered from a slightly acidic peat bog with better bone tissue preserving qualities than highly acidic highland peat bogs (Petska *et al.* 2010). The acidic conditions of high-lying

bogs often result in well-preserved bodies with excellent soft tissue preservation combined with totally decalcified bones (Bennike 2003, 39).

### 3.1.2 Viscera preservation

Our understanding of the long-term survival of viscera (*i.e.* lungs, heart, liver, blood vessels, kidneys and reproductive system) in bog bodies has increased in recent years.

The liver and kidney, the two organs in which the greater part of the volume consists of epithelial cells, are commonly reduced in size, deformed by the pressure of the peat bog layers or unrecognisable, whereas the lungs and intestinal wall (but not its lining epithelium) are usually the best preserved and most recognisable viscera (Aufderheide 2003, 175-176).

In our specific case we were able to confirm the results of the initial macroscopic identification. The paraffin sections of the kidney provided a perfect survey of the kidney tissue, including major characteristic regions such as the cortex and the renal pelvis (fig. 4A). Tubule-like structures were identified in the renal corpuscle (fig. 4B).

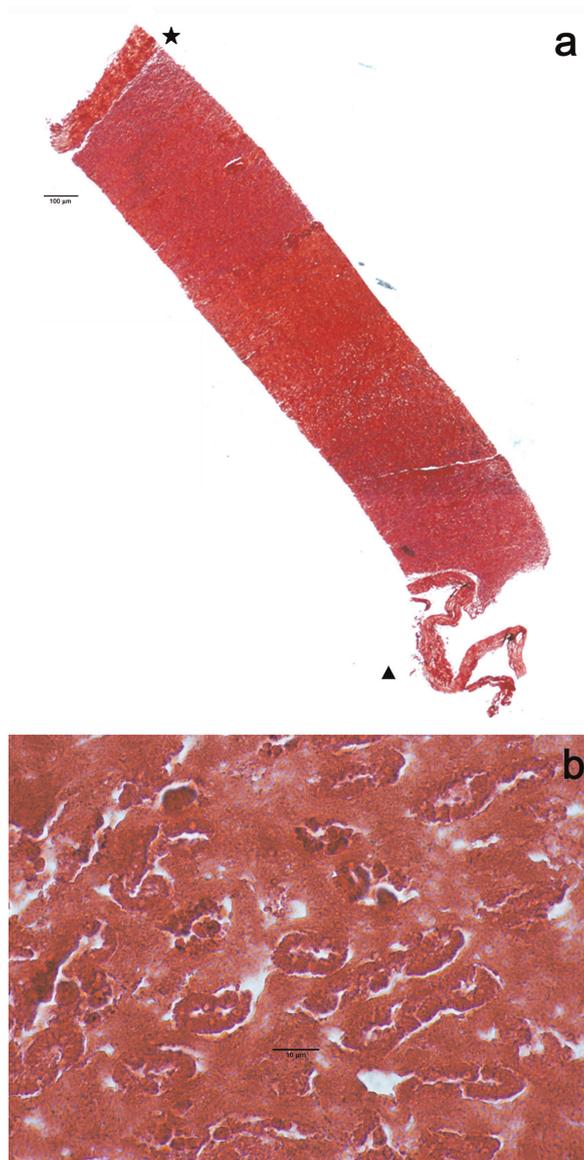


Figure 4 Paraffin section of the kidney. (a) Survey of the kidney comprising three grouped pictures. The kidney capsule (★) and the renal pelvis (▲) are highlighted. Gram stain, bar = 100μm. (b) Detailed view of the kidney cortex with tubular profiles. Gram stain, bar = 10μm.

The liver material appeared to be less well preserved than the kidney sample. Nevertheless, liver parenchyma with polygonal-shaped hepatocytes and connective tissue could still be clearly distinguished in the paraffin sections (fig. 5A).

A relatively large number of eggs of the lancet liver fluke *Dicrocoelium dendriticum* was observed in the liver paraffin sections (fig. 5B). Parasitological findings showing a case of true dicrocoeliasis in a bog body are reported elsewhere (Searcey *et al.* forthcoming).

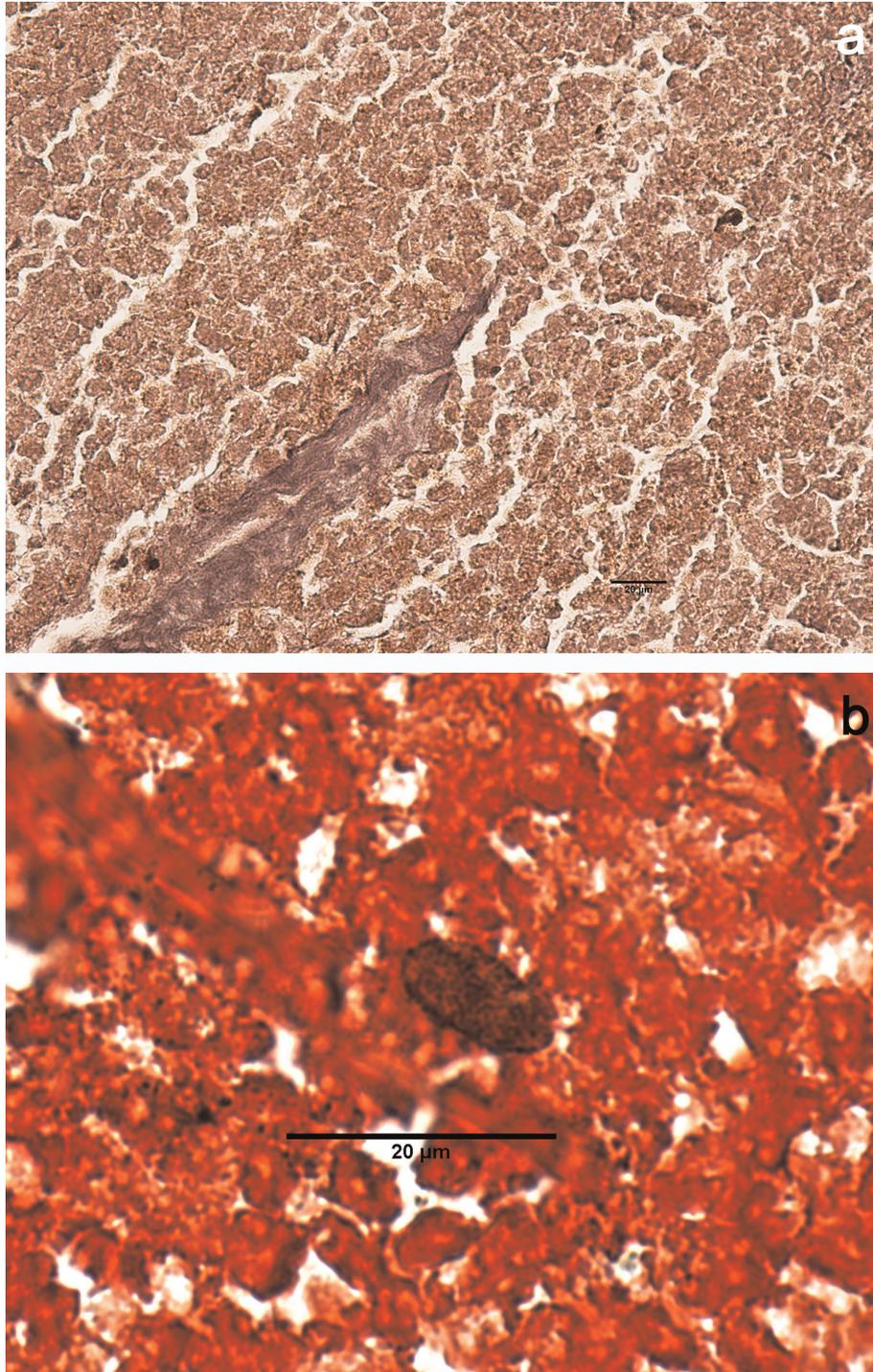


Figure 5 Paraffin section of the liver. (a) Liver parenchyma with polygonal-shaped hepatocytes (brownish) and connective tissue (purple). H&E stain, bar = 20 μm. (b) Egg of *Dicrocoelium dendriticum* embedded in the liver parenchyma. Gram stain, bar = 20 μm.

### 3.2 Tissue preservation analysis by means of atomic force microscopy (AFM)

Of the various forms of connective tissue, collagen fibres are among the most important to survive in bog bodies. They preserve the shape of decalcified bones and the general structure of any remaining parts of the intestinal tract and define the body surface (Brothwell & Gill-Robinson 2002, 126).

AFM images revealed collagen fibrils with periodic banding patterns (figs 6, 7, 8) embedded in the tissue matrix. The average D-period, derived from topographic analysis along the longitudinal axis of several fibrils, was 62.8 nm ( $\pm$  4.2 nm s.d.). The fibrils were unsorted, overlapping at some sites, and formed network-like structures.

Images taken at a greater level of magnification (figs 6, 7, 8) show uninterrupted collagen fibrils, although spherical particles may indicate collagen fragments. The fibril contour structure was very faint. Topographic analysis perpendicular to the longitudinal axis of several fibrils suggested a mean fibril height of 12.2 nm ( $\pm$  3.6 nm s.d.).

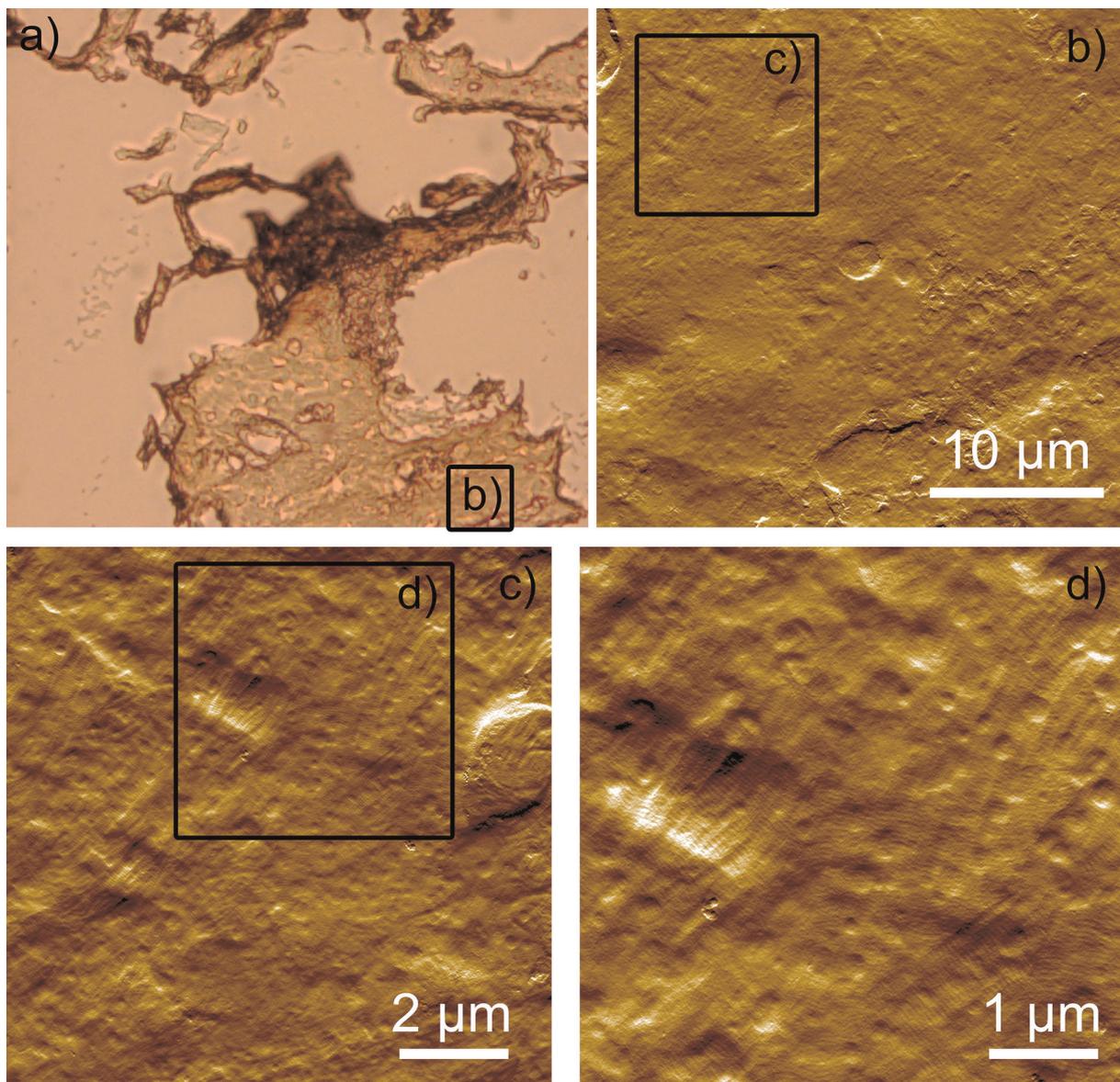


Figure 6 Collagen fibrils within the histological skin sample. (a) Optical microscope image of Zweeloo Woman tissue (magnification 10x). Figures (b) to (d) show magnifying AFM amplitude images of the outlined areas. Figure (d) shows single fibrils with a periodic structure.

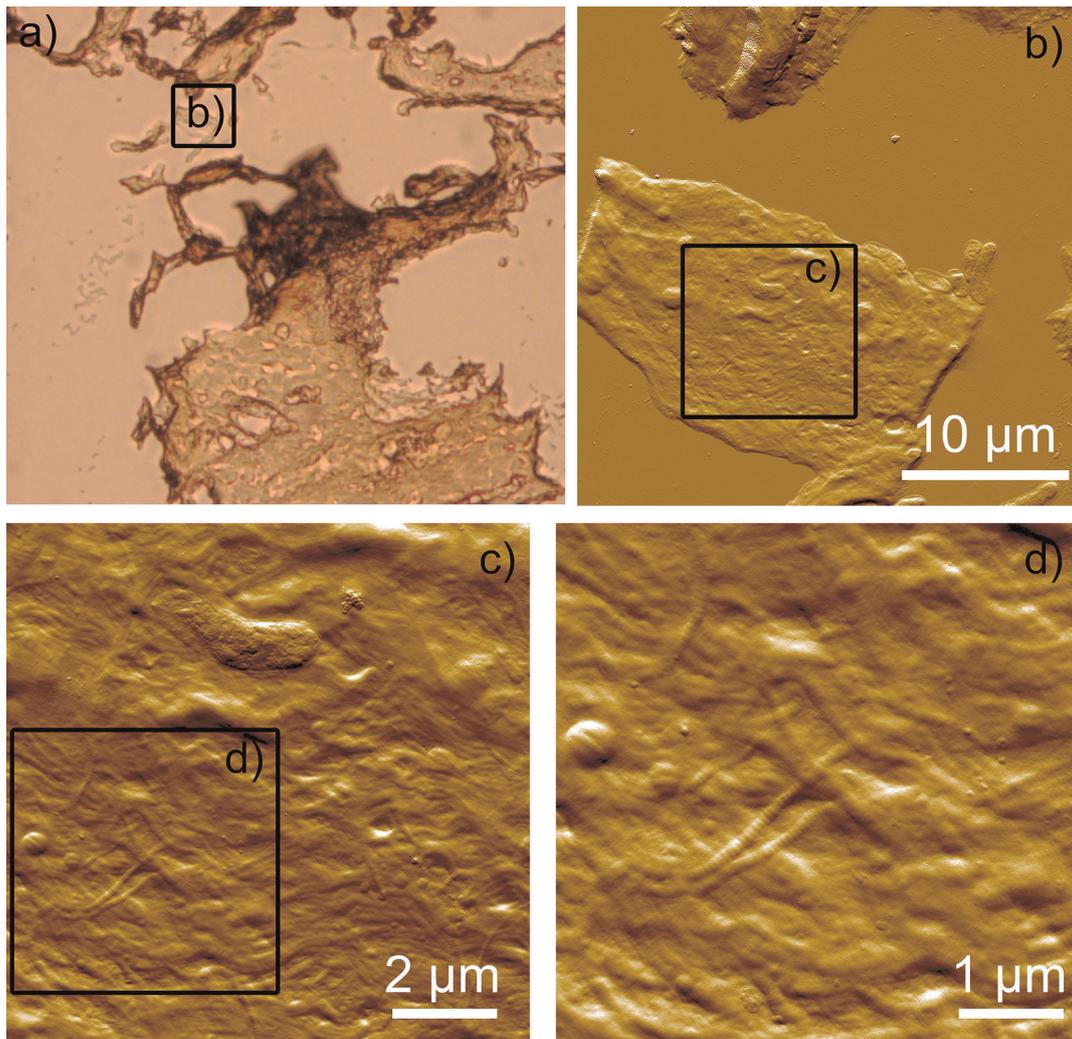


Figure 7 Histological skin sample with collagen fibrils. (a) Zweeloo Woman tissue imaged with an optical microscope (magnification 10x). Figures (b) to (d) show magnifying AFM amplitude images of the outlined areas. Figure (d) shows unsorted, overlapping fibrils in a network-like structure. Each fibril features a periodic substructure.

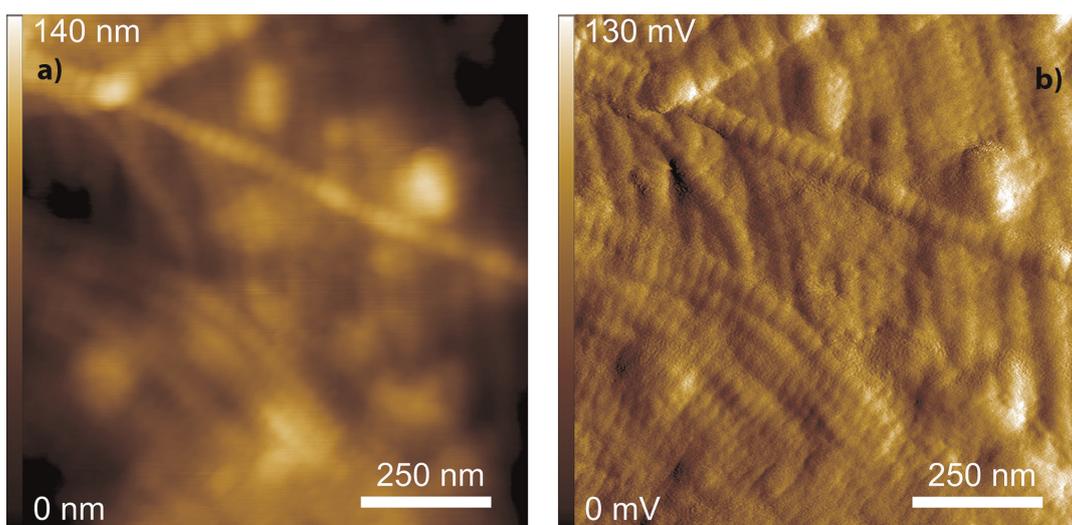


Figure 8 AFM topography and amplitude images of collagen found in the histological skin sample of Zweeloo Woman. Figure (left) shows collagen fibrils (measuring  $2 \times 2 \mu\text{m}$ ) with their characteristic banding pattern. The fibrils form a network-like structure. Some of them overlap one another. The contours of the fibrils are faint. The amplitude image (right) shows the fibril contours in more detail.

Collagen is extremely durable and may survive in mummified tissue for several millennia (Chang *et al.* 2006; Janko *et al.* 2010). This indeed also holds for the structural preservation of the skin collagen of the Zweeloo mummy. As also observed in recent human skin and other mummy skin samples, the collagen fibrils in the skin of Zweeloo Woman were typically arranged in networks or sheet-like structures and showed a periodic banding pattern. Contrary to the results obtained by Stücker *et al.* (2001), who observed well-preserved collagen bundles in the dermis of six bog bodies, our results indicate moderate decomposition of the Zweeloo Woman collagen. The Zweeloo Woman collagen differs considerably in terms of fibril contour and size from the collagen found in recent human skin and other mummies.

High-resolution images taken with the AFM revealed soft outlines of the collagen fibrils, indicating an inferior degree of collagen preservation. The average characteristic banding pattern is  $62.8 \pm 4.2$  nm, which is less than the value of 67 nm reported in the literature. The value is however still within the range of the error margin. The average fibril height of 12.2 nm ( $\pm 3.6$  nm s.d.) is significantly shorter than that of recent skin collagen, which has a diameter ranging from 20 to 100 nm (Fleischmajer *et al.* 1981; Flint *et al.* 1984). This has also been observed in the case of other mummy skin collagen, such as that of the Iceman, whose fibrils were found to have a diameter of 32 nm (Janko *et al.* 2010).

The reduction in fibril height may be caused by decomposition, as observed in previous AFM studies (Paige *et al.* 2002; Bertassoni & Marshall 2009), which revealed the degradation of type I collagen by enzymatic action, e.g. by collagenase or papain-gel. In those studies it was suggested that the enzymes degrade the entire fibrillar structure in a non-specific manner, causing the fibrils to become both shorter and thinner.

An analogous effect may have occurred in the bog and have resulted in slight degradation, hence reduction in size, of the collagen. The differing degree of degradation is most probably due to variations in the composition, in particular the acidity, of the bog governing the preservation and mummification process.

### 3.3 Morphometrical analysis

#### 3.3.1 Pathology: mesomelia

The skull shows clear evidence of a reduction in size (fig. 9) that also affected other parts of the body.

The frontal arc (85 mm) is probably 25 mm smaller than the smallest measurements of female skulls in northern Europe. The sagittal arc (93 mm) is similarly 20 mm smaller than usual for small skulls (Brothwell, personal observations).

These features are very unlikely to indicate microcephaly, a condition which does not normally reduce facial dimensions, but in this case the palate length (30 mm) is 15 mm less than usual.

If we consider ratios of humerus to radius length and femur to tibia, then an arm in a European would normally have a ratio around 1.4: 1.0. In this case it is 2.2: 1.0. The normal ratio of a leg is about 1.2: 1.0; that of Zweeloo Woman is 1.6: 1.0. So the level of reduction in both forearms is considerable, but the length ratio difference in the lower leg is far less, and also shows side-to-side asymmetry (the right leg ratio is 1.28: 1.0).

In the case of congenital reduction in longitudinal segments of the limbs in some forms of mesomelia, forearm reduction may result in ratios of 1.8: 1.0 (personal radiographic observations), which are similar to the Zweeloo ratio. We conclude that the congenital disorder of mesomelia is a possible explanation.

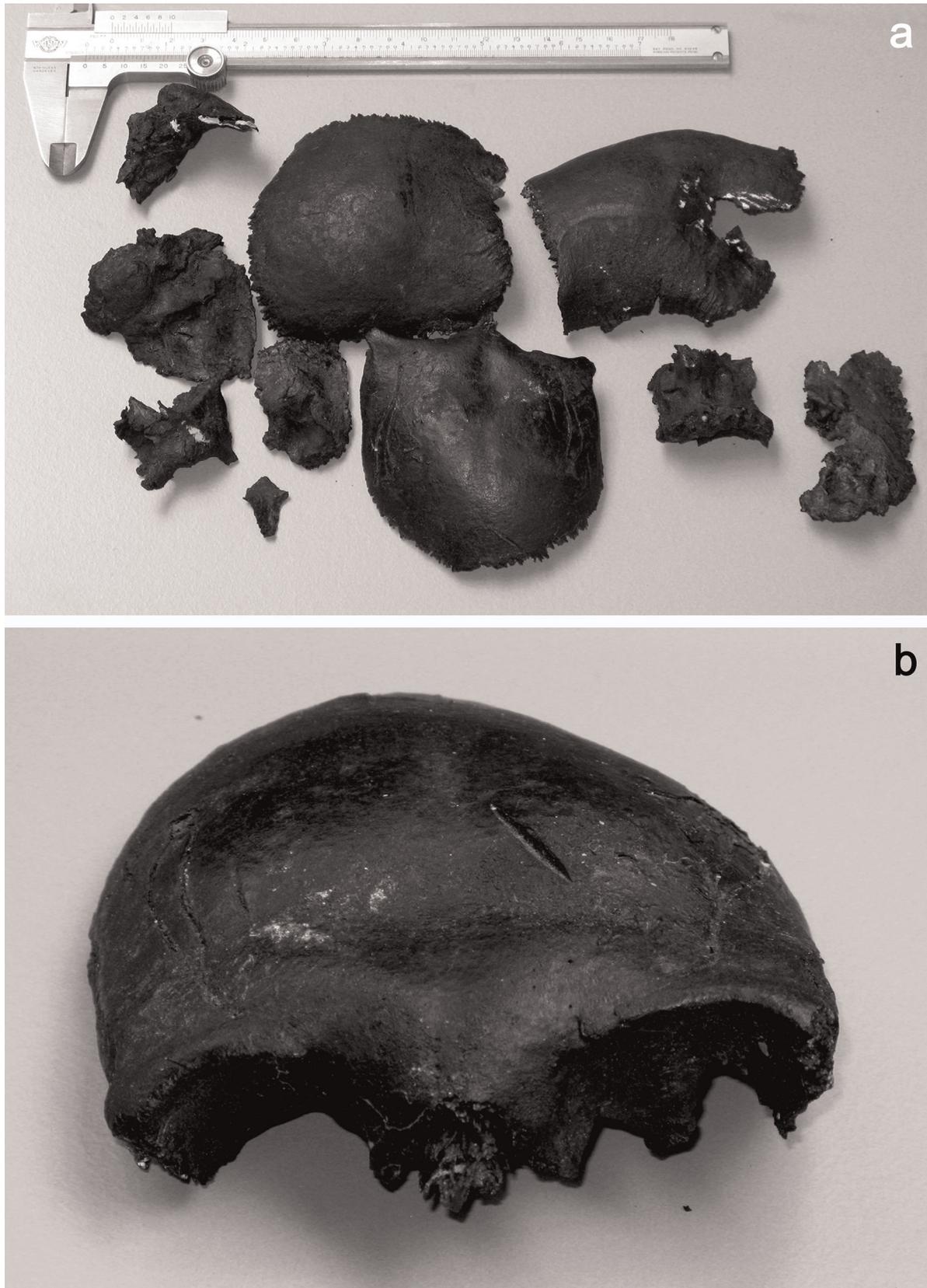


Figure 9 Zweekoo Woman's 'exploded' skull (a) and detail of the frontal bone (b) showing signs of a cut made by a short blade above the left orbit.

### 3.3.2 Stature

Estimation of dimensions on the basis of the outer body surface of the Zweeloo body led to an estimated stature of 155 cm (allowing for head and foot damage). Our measurement coincides with that previously provided by Stoddard (1995), who arrived at an overall stature of about 152 cm.

It should be noted that in a sample of 4,995 British women (Board of Trade 1957), 2,438 (49%) were between 141 and 159 cm, so these are not to be viewed as dwarfed statures. Therefore, Zweeloo Woman could be classified as an example of mesomelia. Approximate dimensions of other body measurements obtained for Zweeloo Woman are presented in figure 10.

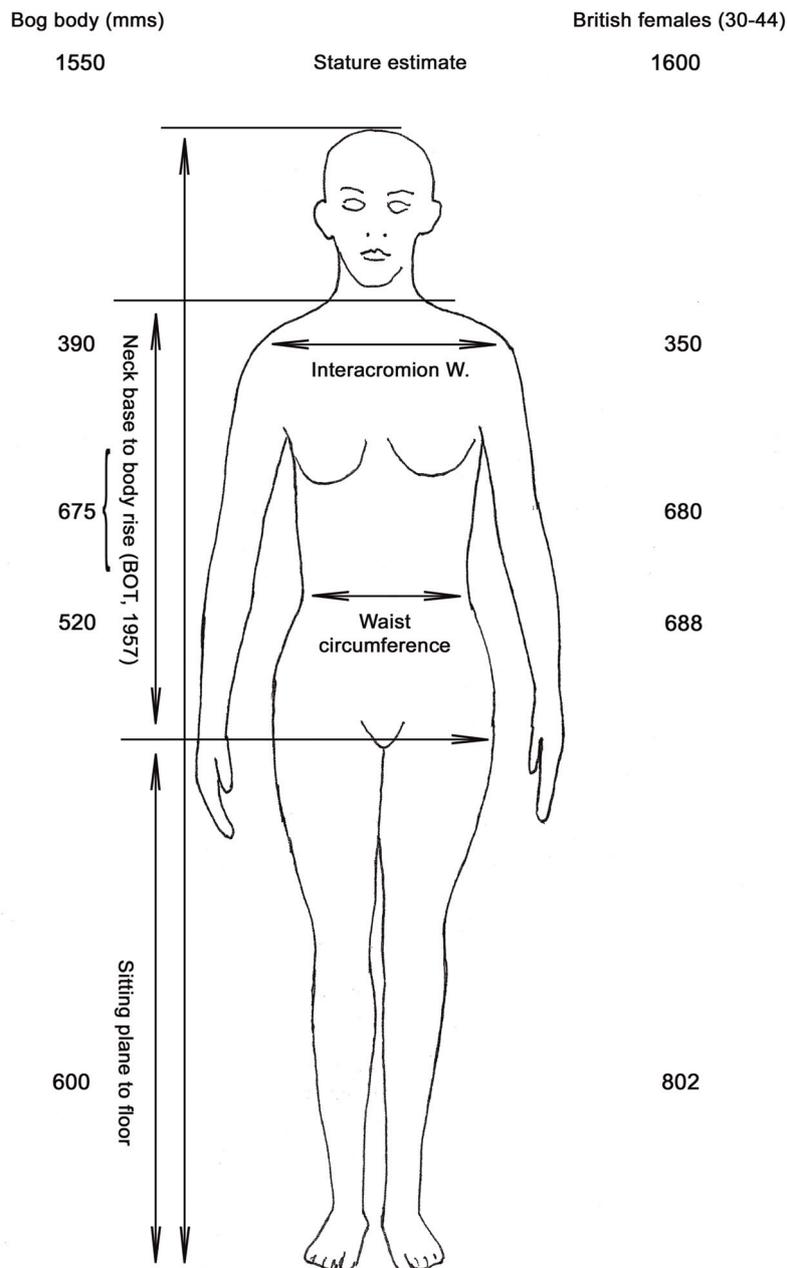


Figure 10 Estimated dimensions of Zweeloo Woman compared with a northern European sample.

The overall length of the trunk of Zweeloo Woman appears to be close to the British average for 30-44 year olds (Board of Trade 1957). The leg length, however, seems to be shorter (with due allowance for the damaged feet). The interacromion width of the Zweeloo body may be above average, but the waist circumference appears to be slightly smaller than the modern European mean. However, neither difference is significant.

	Length* in cm (RB, DPM)	Length in cm (DB)	Mean
Right humerus	26.3	26.6	26.45
Left humerus	#	#	#
Right radius	12.1	12.5	12.3
Left radius	11.8	12.04	12.1
Right ulna	#	#	#
Left ulna	12.7	13	12.82
Right femur	32.6	(35.5)**	34.05
Left femur	#	#	#
Left tibia	21	21.8	21.4
Right fibula	21.6	#	#
Left fibula	18.1	#	#

# Fractured bones for which measurement could not be performed

\* Lengths defined in Brothwell (1981)

\*\* Estimated with curvature corrected

*Table 1 Measured lengths of bones (in cm) of Zweeloo Woman*

The stature estimated on the basis of the bones of Zweeloo Woman ranges from 130 to 135 cm when the humerus, femur and tibia are included in the calculation, and from 137 cm to 141 cm when the tibia is excluded (Table 2).

Index*	Minimum	Maximum	Mean	Comparative data <sup>x</sup>
Humero/radial	46	47	46.5	76.6 ± 1.8
Femuro/humeral	80.1	74.4	77.7	71.2 ± 1.3
Femuro/tibial	65.2	62.1	62.9	81.6 ± 1.7

\*Indexes after Martin (1928)

<sup>x</sup>Mean of 138 central European populations with a total of 14,730 individuals, after Siegmund (2010)

*Table 2 Stature of Zweeloo Woman estimated on the basis of the bones*

The Pearson method was considered the most appropriate among the estimation methods, since the employed reference series corresponds most closely to European populations in terms of body proportions (Siegmund 2010, 73-76) and Pearson considers all the long bones in the regression. In contrast, Trotter & Gleser (1952) consider only the tibia, and when the tibia is not available they use the femur. This calculation method would have led to distortions in the case of Zweeloo Woman due to the short length of the tibia and the curvature of the femur.

Comparison with a large dataset of an archaeological population from central Europe contemporary with Zweeloo Woman shows that she was significantly shorter than the mean (153.3 cm) of the female population of this period (Siegmund 2010, 83). With due allowance for the standard deviation and the interquartile range, 96% of the female population of this period ranged from 144.3 to 162.3 cm. Taking into account the bone shrinkage commonly found in bog bodies, the stature of Zweeloo Woman could be considered short, but within normal limits.

### 3.3.3 Body proportions

Besides the stature, the body proportions were also calculated according to Martin (1928, 1067) (Table 3) to enable us to evaluate the unusual proportions of Zweeloo Woman. The FHI and FTI are both significantly smaller than those of central European populations, which vary (Siegmund 2010, 62, 64). The HRI is extremely low due to the abnormal shortening of the radius.

Stature estimation with tibia		Stature in cm		
Stature estimation method		Minimum	Maximum	Mean
Pearson (1899)		133.6	137.1	135.3
Trotter/Gleser, white (1952)		127.7	132.8	130.3
Trotter/Gleser, negro (1952)		127.3	131.9	129.6
Mean of Pearson, Trotter/Gleser		129.52	133.94	131.73

Stature estimation without tibia		Stature in cm		
Stature estimation method		Minimum	Maximum	Mean
Pearson (1899)		139.4	143	141.3
Trotter/Gleser, white (1952)		134.6	141.8	138.2
Trotter/Gleser, negro (1952)		134.1	140.7	137.4
Mean of Pearson, Trotter/Gleser		136	141.8	138.9

Table 3 Body proportion estimation of Zweeloo Woman

### 3.3.4 Body Mass Index

The Body Mass Index (BMI) of Zweeloo Woman was calculated on the basis of the estimated stature and the femur head diameter using the formula of Auerbach & Ruff (2004) [which is actually a mean of the formulas of Ruff *et al.* (1991), McHenry (1992) and Grine *et al.* (1995) combined with Pearson's method]. The BMI value of Zweeloo Woman is normal in comparison with present-day data provided by the World Health Organisation (2006), according to which values from 18.50 to 24.99 fall within the normal weight range (Table 4).

		Stature estimation after Pearson	Body Mass estimation (values in kilogram)				BMI
			after Ruff <i>et al.</i> 1991	after McHenry 1992	after Grine <i>et al.</i> 1995	mean (after Auerbach & Ruff 2004)	
without tibia	min	139.4	40.46	33.99	38.34	37.60	19.34
	max	143.0	40.46	33.99	38.34	37.60	18.39
	mean	141.2	40.46	33.99	38.34	37.60	18.86
with tibia	min	133.6	40.46	33.99	38.34	37.60	21.07
	max	137.1	40.46	33.99	38.34	37.60	20.01
	mean	135.3	40.46	33.99	38.34	37.60	20.53

Table 4 Body Mass Index estimation of Zweeloo Woman

## 3.4 Radiographic analysis by means of CT scanning

### 3.4.1 Sex

The sex estimation was based on the pelvis and parts of the skull (White & Folkens 2005, 385-398). The following bones were entirely preserved: frontal bone, right and left parietals, a fragment of the left temporal bone and a fragment of the upper jaw. The reduced dimensions of the

skull bones are due to post-mortem shrinkage and not microcephaly. True microcephaly does not result in such a pronounced reduction of the face.

The very small mastoid process of the skull and the shape of the pelvis (wider greater sciatic notches, longer pubic portion of the *os coxae*, larger subpubic angle, more elevated auricular surface) indicate that the individual was female.

### 3.4.2 Age at death

The age at death was tentatively inferred from various observations. Epiphyseal union in the long bones, pelvis and scapula indicates a minimum age of 25 years, while the morphology of the pubic symphysis points to an age between 35 and 50 years (Leopold & Schäfer 1998, 304-308; Byers 2005, 223-232).

### 3.4.3 Cut marks

There are at least 21 cut marks on the bones. Most of the cuts are short and were clearly made with a sharp blade. There is no evidence of bone reaction to these injuries, and there are no equivalent injuries on the body surface. The distribution of these injuries is shown in figure 11.

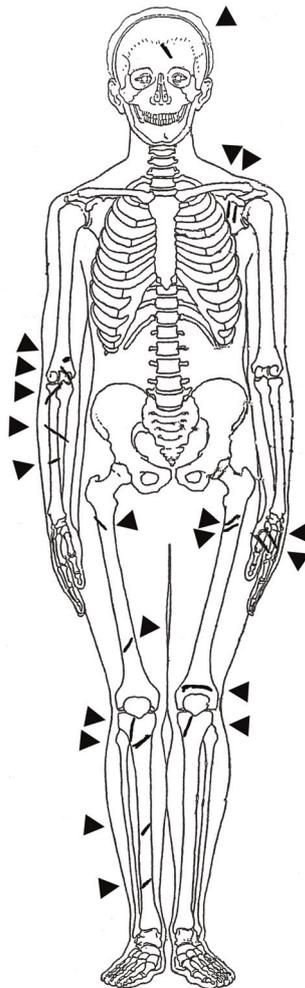


Figure 11 Distribution of cut marks on the Zweeloo skeleton.

Briefly, the locations of these are as follows: (1) frontal, above the left orbit, (2) two on the inner aspect of the scapula blade, (3) a left hand phalanx and proximal phalanx of the left thumb, (4) the left femur below the trochanters and at the distal femur, (5) at the proximal articular end of the left tibia, (6) two at the distal end of the right humerus, (7) three on the right radius and one on the right ulna, (8) two on the right femur, in the upper and lower thirds of the shaft, (9) two at the proximal end of the right tibia and two more along the shaft. The cause of the damage is unclear, as there is no evidence of external body trauma, except possibly to the posterior aspect of the left shoulder, on the outer skin surface. It remains to be determined whether the cut marks were formed during the excavation or during the conservation of the remains.

#### 4 Pseudopathology (deformation) and other abnormalities

Excluding the various cuts, and the possible congenital anomaly under discussion, a number of other pathological conditions were noted. The pelvis shows some long deep grooves on the left iliac blade which are probably due to post-mortem shrinkage and distortion. Two upper thoracic and four lumbar vertebrae show varying degrees of Schmorl's nodes. They were originally thought to have no clinical significance, but may well be associated with osteoarthritic development, back stress and pain (Aufderheide & Rodríguez Martín 1998, 96-97; Capasso *et al.* 1999, 38).

The postcranial skeleton is almost entirely preserved, although a marked difference in size is observable between some bones of the left side of the body and their counterparts on the right side (i.e. the pelvis, the heel bones and the femora). Since these observations are not in accordance with any pathological disorder, post-mortem deformation is suggested as the main cause of these differences.

If asymmetry is indeed a post-mortem development, the specific shortening of the forearms and lower legs should possibly be classified as a pathological phenomenon known as dyschondrosteosis or Léri-Weill syndrome.

##### 4.1 Possible anomaly of the forearms and lower limbs

Both the proximal and the distal epiphyses of the right humerus and the preserved distal epiphysis of the left humerus are normal in shape and structure. The right and left forearms show partially deformed and underdeveloped epiphyses. The left and right radii are shorter than normal and moderately bowed.

The left femur is fractured and a small radiopaque body (1.5-2 cm, HF density between 1.000 and 2.000) is observable at the fracture edge of its distal epiphysis. This structure seems to be an intrusive stone (fig. 12).

Both the right femur and the right fibula show marked bowing. There are signs of *coxa valga*. The left leg appears to be shorter than its counterpart.

Both lower legs appear to be underdeveloped in comparison with the femora. This difference is however less evident than that observed in the forearms.

Proximal portions of the limbs, hands and feet show no abnormality of size, though there is possible evidence of reactive change in the bones of the feet caused by some degree of abnormal gait. There is no evidence of nutritional deficiency or infectious illness, and no other part of the skeleton appears to be affected.

The X-ray features of dyschondrosteosis in living patients and in dry bones are based on a spectrum of manifestations. The features typical of DCS patients that are observable in Zweeloo Woman include: 1) shortening of the radius extremity with hypodevelopment of both proximal and distal epiphyses as compared with the normal standard for age and in relation to the

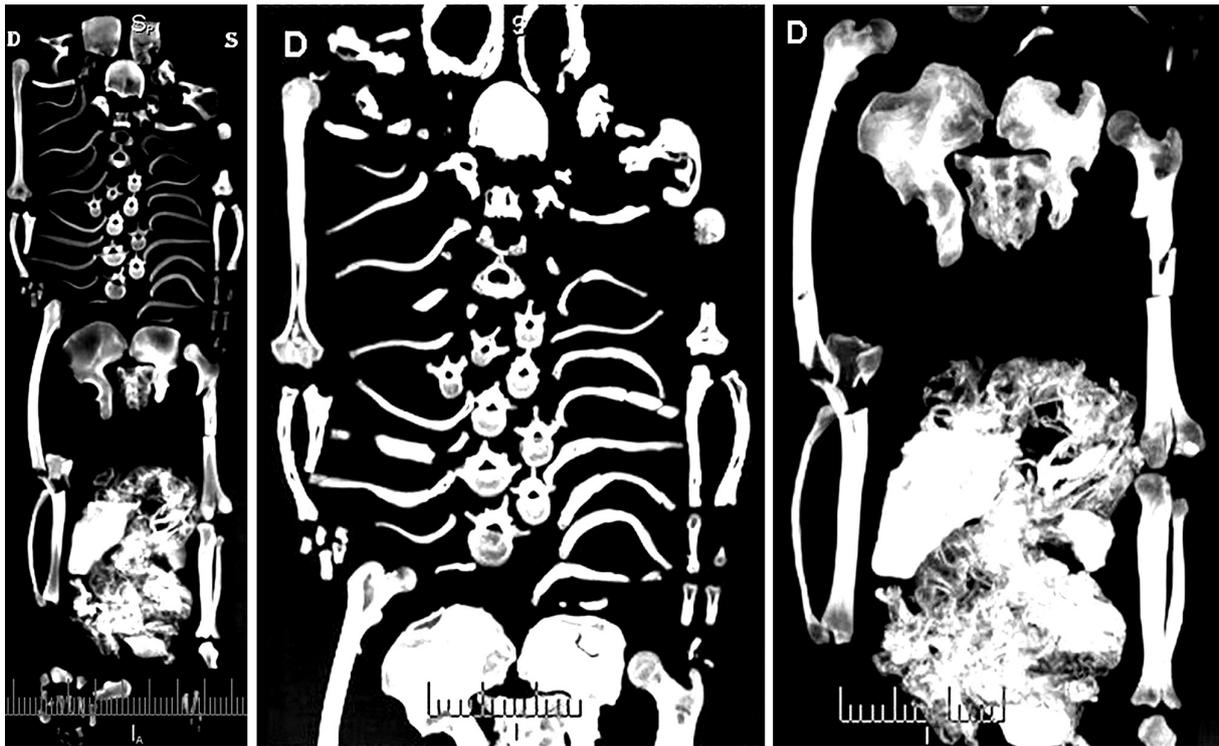


Figure 12 Results of a CT scan performed on Zweeloo Woman's skeletal remains. The viscera were positioned between the lower limbs.

size of the other bones; 2) double (lateral and dorsal) bowing of the radius which involves the entire diaphysis but is more marked at the distal end (Madelung's deformity; Langer 1965); 3) shortening of the tibia in relation to the femur, which results in mild dwarfism. This pattern of shortening in the middle segment of extremities is referred to as mesomelia (Langer 1965).

The skull, the spine and the pelvis are radiologically normal in all cases described in the literature in which these bones have been studied, those of Zweeloo Woman included.

Surprisingly few bog bodies have been reported as showing anomalies and, where observed, the disease is mostly associated with adult individuals (Brothwell & Gill-Robinson, 2002, 120).

Following Lynnerup (2010, 444) "acid bog diagenetics mean that bone will be demineralised, become pliable and, upon subsequent excavation and drying out, also shrink and warp. This means that under these conditions, etiological attribution of pathology and trauma lack certainty".

The greatest uncertainty in the case of Zweeloo Woman is essentially her 'true' stature. The great discrepancy between the value of 170 cm quoted shortly after her excavation (1952) and the values of 152 and 155 obtained in 1995 and 2009-2011 is clearly an issue.

No written records of the stature measurements carried out at the time of exhumation are available. The only scientific information is that provided by Stoddart, who interpreted the asymmetry identified between one side of the body and its counterpart as due to shrinkage.

The data obtained by means of AFM and histology provide evidence of slight degradation of collagen fibrils and moderate bone demineralisation, implying shrinkage. The possible diagnosis of Léri-Weill dyschondrosteosis should however not be totally rejected.

Since the taphonomic impact of the acidic peat bog environment on human remains is still not well understood (Janaway *et al.* 2003, 56-59; Gill-Frerking & Healey 2011, 69-74), the possibilities of taphonomy and pseudopathology must always be considered (Gill-Robinson 2003, 46). Similarly, bone abnormalities should not always be assumed to be due to bog taphonomy.

The possibility of skeletal disorders pre-existing the taphonomic process should also be considered.

To sum up, the bone morphology and radiological findings indicate a possible case of Léri-Weill dyschondrosteosis.

#### 4.2 *Differential diagnosis*

Differential diagnosis should include other deformities of the distal forearm (Langer 1965; Leiber & Olbrich 1981, 1135). Post-traumatic fusion of the ulnar aspect of the distal radial epiphysis may closely simulate the deformity of dyschondrosteosis, but Zweekoo Woman does not show any signs of such traumatic lesions.

Less commonly, infection may result in a similar deformity. The deformity is almost always unilateral in such cases. As is evident from all the published cases, dyschondrosteosis may be asymmetric as regards involvement, but, invariably, the criteria of dyschondrosteosis are radiologically observable, in both the forearms and the wrist regions (Langer 1965).

This is not a case of hypopituitary dwarfism, since it is not a generalised developmental failure. In achondroplasia, the most common form of skeletal dysplasia, the limbs are all shortened – the femur the most, then the humerus, then the bones of the lower legs and forearms. Adult stature rarely exceeds 140 cm (Aufderheide & Rodríguez Martín 1998, 360).

Achondrogenesis and thanatomorphic and camptomelic dysplasia are excluded from a differential diagnosis, because affected people die stillborn or at birth, and within the first year, respectively. Pseudoachondroplasia is also excluded as it is prominent at the hips (femora) and shoulder (humeri), with irregular epiphyses and widened metaphyses resulting in premature degenerative joint disease (Aufderheide & Rodríguez Martín 1998, 360-361).

Similarly, it is not a phocomelic type of dwarfism, since only a specific segment of each limb is involved and the skull is unaffected. The normal hands and feet argue against an effect of the 'thalidomide injury' type, for example following a viral infection. An 'anti-Marfan' type of syndrome can also be excluded, because the equivalent segment is involved in each limb, and the skull, hands and feet are spared (Langer 1965; Stoddart 1995, 9).

#### 4.3 *Genetic and clinical aspects of Léri-Weill dyschondrosteosis*

Léri-Weill dyschondrosteosis (LWD) is a dominantly inherited skeletal dysplasia marked by disproportionate short stature and the characteristic Madelung's wrist deformity. LWD is inherited in a pseudo-autosomal dominant manner with each child of an affected individual having a 50% chance of inheriting the mutation. Prevalence is unknown.

Short stature is present from birth with mesomelic shortening of the limbs (shortening of middle segments of the forearms and lower legs). Madelung's deformity may only be detected at puberty. The wrist deformity is bilateral and is characterised by shortened and bowed radii and ulnae leading to dorsal dislocation of the distal ulna and limited mobility of the wrist and elbow (Langer 1965).

LWD was first described by Léri and Weill in 1929 in a French paper and since then occasional reports have appeared in the French, German and Spanish literature (Léri & Weill 1929; Langer 1965). Although the disorder occurs in both sexes, it is usually more severe in females, possibly due to sex differences in oestrogen levels (Lichtenstein *et al.* 1980).

However, pubertal development and fertility are generally normal in both sexes with the disorder (Ross *et al.* 2003). Intelligence is normal. In around 70% of cases, LWD is caused by haploinsufficiency of the short stature homeobox (SHOX) gene, which maps to the pseudoautosomal region 1 (PAR1) of the sex chromosome (Xp22.23 and Yp11.32) (Belin *et al.* 1998; Shears

*et al.* 1998; Ross *et al.* 2001, 2003; Huber *et al.* 2001; Grigelioniene *et al.* 2000; Benito-Sanz *et al.* 2005).

Haploinsufficiency results from heterozygous mutations and deletion of SHOX, or of the downstream PAR1 (where SHOX enhancer elements are located). The molecular defect remains unknown in the remaining 30% of LDW cases. SHOX-associated LDW is part of a spectrum of disorders (ranging from the most severe Langer mesomelic dysplasia (LMD) to LDW, isolated Madelung's deformity and so-called idiopathic short stature) all associated with SHOX/PAR1 abnormalities. The prevalence of SHOX/PAR1 mutation is estimated at 1/1000.

Diagnosis of suspected LDW in the Zweekoo Woman on the basis of the clinical and the radiologic findings will be attempted by means of molecular analysis in a follow-up study. However, we are aware of the fact that in some cases in which the degree of preservation may seem remarkably good, the chances of aDNA having survived in these acid wetland bodies will be very low.

## 5 Other archaeological studies

Several cases of probable skeletal dysplasia in pre-modern societies have been described. The principal cases discussed in the literature range from the Upper Palaeolithic to the Middle Ages, but none involve a bog body. Those principal cases are listed below.

- An Upper Palaeolithic male individual from Italy represents the earliest case of dwarfism associated with Madelung's deformity. Other indications in this skeleton tell us that he did not suffer from DCS but a different form of dwarfism, an acromesomelic dysplasia, which is much more severe than DCS. The stature of this individual was estimated at only 1.0 to 1.30 m (Fruyer *et al.* 1987).
- A 2nd-3rd-century AD young female from the Talayotic site of 'S. Illot de Porros' in Mallorca, Spain (Campillo & Malgosa 1991) with evidence of bilateral brachymelia with bowing of the radius and posterior dislocation of the ulna consistent with probable Madelung's deformity.
- A mature adult male (45-50 years), the Donori man, dating from 2800 BC from Nuragic in Sardinia. The main pathological features affected the forearms, which were both characterised by marked shortness and morphological alteration of the distal epiphyses. Some major changes involved the radii, whose diaphyses were shortened and strongly bowed. But as the lower legs had not survived, a diagnosis of mesomelia could not be made and an aetiology of DCS could not be stated with certainty (Canci *et al.* 2002).
- A 20-25 year old male skeleton dating from the 2nd-3rd century AD from Gloucester, England. The most significant abnormalities were Madelung's deformity affecting the right radius (the only one to have survived sufficiently well to allow examination), an abnormal right humerus, curved and shorter ulnae and mesomelia, suggesting a probable diagnosis of dyschondrosteosis (Waldron 2000).
- A 60 year old female skeleton dating from the 6th century AD from Geneva, Switzerland, showed the forearm changes and may be an example of DCS (Kauffman *et al.* 1979).
- A 7th century male aged between 18 and 20 years from the Pieve di Pava, Siena, Italy, whose skeleton reveals a stature of about 150 cm with short, bowed radii, bilateral deformation of the proximal and distal epiphyses of the radii and ulnae, bilateral bowing and shortening of the tibiae and bilateral agenesis of the fibulae. These deformations are suggestive of Langer syndrome (Mongelli *et al.* 2010).
- A 9th-11th-century Anglo-Saxon female from Black Gate cemetery, Newcastle-upon-Tyne, England. The age at death was estimated at between 35 and 44 years. This skeleton revealed deformity of both forearms and shortened stature due to reduced tibial length, which were considered indicative of dyschondrosteosis (Cummings & Rega 2008).

- An 11th-13th-century adult male skeleton from the Hispano-Muslim necropolis of San Nicolás de Murcia, Spain. The deformities of the forearms are quite similar to those of the Donoriman, especially those concerning the medial distal epiphysis of the radius (Campo *et al.* 1996).
- A medieval elderly male from Saint Gregory's Priory, Canterbury, Kent, England. In this case there is no evidence of dysplasia. The right radius and ulna were shortened due to premature fusion of the distal growth plate. The radius displayed lateral and dorsal bowing and the ulna was dislocated posteriorly. The unilateral presentation, shortening of the ulna and atrophy of the hand bones suggest a post-traumatic rather than an idiopathic form of the Madelung's deformity. The normal length of the lower legs is an argument against a diagnosis of dyschondrosteosis and Madelung's deformity associated with mesomelic dysplasia (Anderson & Carter 1995).

Only three cases of probable or indicative DSC have been described so far. Zweeloo Woman's skeletal remains (upper and lower limbs) show marked dysplasia consistent with a mesomelic abnormality and provide possible evidence of a Léri-Weill syndrome in a bog body.

## 6 Possible archaeological significance

### 6.1 *Zweeloo Woman today (2011)*

Zweeloo Woman was an adult female who lived some time between the late 1st and early 3rd century AD. She died at an age between about 35 and 50 years. Under normal circumstances a local woman would have been cremated and the remains buried at the local cemetery. Her final resting place, however, was a pit in a bog in which she was disposed of probably naked in a somewhat foetal position. Her last meal most probably consisted of a gruel based mainly on millet, perhaps also containing the blackberries whose pips indicate a death in late summer-early autumn.

The new series of investigations carried out on this bog body confirm the previous tentative diagnosis of dyschondrosteosis as the cause of the disability that affected Zweeloo Woman during her life. Zweeloo Woman was of short stature, and may have appeared somewhat bizarre to others due to her physical deformity. Whether she died an unnatural death it is not known with certainty because no unambiguous signs of external violence are visible on her skin, but the fact that her hair was cut not long before her death may indicate that her death was not natural (Asingh 2007, 293). Her unusual burial may well be connected with her deformity having afforded her a specific place in society. It is very tempting to associate her physical appearance with her unusual treatment, i.e. being buried in a bog. There are other bog bodies showing body defects or anomalies: Bolkilde Man was crippled, the Døjringe Men had defects in their upper arms, Yde Girl suffered from scoliosis and Lindow Man (III) had a hand with an extra digit, to name only a few examples spanning a period of several millennia (see Van der Sanden 1996, 138-143). However, because our dataset is so small and deficient, the possibility that the physical condition played a role in the selection process by which people were earmarked for an unusual death/burial in a bog will probably continue to remain an attractive hypothesis.

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## Glossary of the employed medical terms & definitions

**Dicrocoeliasis** = hepatic fascioliasis due to infection with *Dicrocoelium dendriticum*.

**Dyschondrosteosis** = a familial bone dysplasia characterised by bowing of the radius, dorsal dislocation of the distal ulna and proximal carpal bones, and mesomelic dwarfism. Also called Léri-Weill syndrome.

**Dysplasia** = an abnormality of development; in pathology, alteration in size, shape and organisation of adult cells.

**Mesomelia** = a condition in which the forearms and lower legs are abnormally short.

**Micromelia** = an abnormal shortness or smallness of limbs. Also called nanomelia.

## Notes

1. Now known as *Groninger Instituut voor Archeologie* / Groningen Institute of Archaeology.
2. Information provided to one of the authors (WVDS) by F. Kuipers and J. Warmolts, Zweeloo, on February 5 2010.
3. By the time of the discovery the peat bog no longer had its original shape. Topographic maps from the 1920s show that Juffersveen had by then 'broken up' into smaller parts.
4. One of the authors (WVDS) searched for such documents in the late 1980s.
5. Calibrated using Wincal25.
6. In a letter to one of the authors (WVDS) he writes that the woman will have been about 1.52 m tall.

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