Hypermobility in reproduction

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Peripartum pelvic pain (PPPP) syndrome is thought to be caused by decreased stability of the pelvic girdle, caused by increased relaxin levels during pregnancy. The resulting hypermobility (HM) may result in back pain, symphysiodynia and waddling gait. The hypothesis that pre-existent hypermobility is a major cause of PPPP was investigated.

Surveys of HM and HM plus PPPP were conducted in different populations in several countries. The prevalence of HM in non-pregnant nulliparous women ranged between 10% and 39%; cross-sectional studies in pregnant women showed a prevalence between 5% and 19%. Significant correlations between HM and PPPPS were found in the Dutch subjects only.

There seems to be a genetic difference between populations with regard to the development of PPPP in HM pregnant women. Pregnancy is the main trigger causing development of benign joint hypermobility syndrome (BJHS) in later life.

The symptoms of peripartum pelvic pain (PPPP), also called ‘pelvic insufficiency during pregnancy’, ‘pelvic girdle relaxation’ or ‘symphysiolysis/symphysiodynia’, can be described as pain, which develops during pregnancy, in the symphysis, sacro-iliac (SI) joint, os coccygis, ossa pubis, tubercula pubica and tubera ischiadica with or without radiation to the posterior side of the upper leg. The syndrome can be accompanied by a typical waddling gait. Symptoms usually start before 20 weeks’ gestation and are more pronounced in multigravidas. All the sensations tend to worsen during pregnancy, even to a stage of complete incapacitation. The frequency in Western Europe is estimated at between 8 and 19/1000 deliveries, with a rate of recurrence in a subsequent pregnancy of 41 - 85%.1-2 Significant correlations between HM and PPPPS were found in the Dutch subjects only.

Back pain occurs in about 50% of all pregnant women.2,4 Chronic back pain in older women started in former pregnancies in 10 - 28% of cases, suggesting that pregnancy itself is the main risk factor for the development of nonspecific back pain.2,6

Pathogenesis of PPPP

Hormonal effects

Increased relaxin (Rlx) levels during pregnancy were thought to cause PPPP,7 but other studies showed no correlation between Rlx levels and the severity of symptoms of PPPP.8-12 A survey of 518 women who had suffered from PPPP showed that 72% experienced premenstrual relapse of symptoms, and that 85% had a recurrence of PPPP in a subsequent pregnancy.1

Mechanical factors

• PPPP was associated with high-weight babies in one study1 but not in other ones.13-15 There was an increased chance of instrumental delivery, reflecting mechanical problems and higher birth weight, but not an increased caesarean section rate.1

• Pelvic stability is maintained by the anatomy of the SI joint, so-called ‘form closure’, and compression forces of ligaments and muscles, the ‘force closure’.17 Larger movements in the SI joint due to increased Rlx levels during pregnancy cause decreased stability of the pelvic girdle.

• Increased mobility between the pubic bones resulted in an average 6 mm symphyseal movement in puerperal women with PPPP, compared with 2 mm in women without.16,18 Increased joint laxity in pregnancy may cause a vicious cycle of pain and fatigue due to decreased proprioception, decreased muscle function and stability, and impaired load transfer from trunk to legs, resulting in ‘catching of the legs’ (i.e. experiencing difficulty in moving one or both legs when walking, resulting in more pain, fatigue, etc.).17

• Radiographs taken with the patient standing on one leg, alternating the symptomatic and non-
symptomatic sides, showed a significantly larger caudal shift of the pubic bones on the affected side.\textsuperscript{18,19} The typical waddling gait in PPPP patients may be explained by insufficiency of the abductor muscles and increased body weight, resulting in inability to keep the pelvis horizontal during the one-leg stage of a walking stride. The patient will place the body’s centre of gravity above the hips of the weight-bearing leg (Trendelenburg sign).

Increased awareness

The sudden appearance of PPPP could also be due to increased awareness of this syndrome on the part of patients.

Intrinsic hypermobility

Larsson’s 1993 publication\textsuperscript{20} on joint hypermobility in musicians who experienced increasing joint problems during their professional lives led to the hypothesis that pre-existent hypermobility (HM) during pregnancy could be the main cause of PPPP.

Peripartum pelvic pain score (PPPS)

The PPPS was developed to measure the symptoms of PPPP objectively. The scoring system (Table I) comprises maxima of 6 points for the active straight leg raising test,\textsuperscript{20} 3 points for the femoral flexion test\textsuperscript{21} and 6 points for the adductor test.\textsuperscript{22} The maximum total PPPP score is 15/15, and the cut-off point for severe PPPP during pregnancy has been set at > 12/15.

Active straight leg raising test (ASLR).\textsuperscript{23} The patient raises one then the other hyperextended leg to a height of 10 cm. If there is too much mobility in the symphysis, the musculi recti femoris will pull the two pelvic bones into a painful frictional symphyseal rotation.

Femoral flexion test (FFT) or posterior pelvic pain provocation test.\textsuperscript{22} The upper leg is flexed perpendicularly in the hip joint. Gentle pressure is put on the ipsilateral knee while the other hand fixes the contralateral hip. This is a good measure of SI involvement. The pain must be localised in the ipsilateral SI joint or buttock.

Active testing of adductor strength (ADD).\textsuperscript{23} It has been shown that the average muscle power of the adductors amounts to 222 (±60) Newton (N), whereas most women with PPPP score below 150 N. The patient is positioned with the feet flat and the knee joint at a 90° angle. The force is measured by placing a dynamometer between the knees and instructing the subject to squeeze the hand for 5 seconds.

Hypermobility

History

The painter Matthias Grünewald (1450 - 1528) produced the Heller Retable (Frankfurt, Städliches Kunstinstitut) depicting Saint Cyriac (fecit 1511 - 12) and a kneeling young woman: both show hyperextension of thumbs or little fingers.

In circuses all over the world contortionists earn their living twisting their bodies into the most impossible postures, which they are able to do because of their extreme generalised HM or hyperlaxity.

The nomadic Scythes were formidable archers on horseback. About 700 BC they conquered territory from southern Russia to Mongolia. The Greek physician Hippocrates (460 - 377 BC) speculated that the Scythes could maintain their large territory because of their extreme flexibility: they rode horses bareback and stayed on by wrapping their legs around the horse’s belly. Eventually, however, they could not handle large bows any more because of frequent (sub-)luxations of the shoulder joints.\textsuperscript{24} The Scythes disappeared from history and were integrated into the population of Mesopotamia. A study among Iraqi students revealed the highest HM frequency in the world: 38% for females and 25% for males.\textsuperscript{25} HM, initially an advantage for the Scythes, eventually became their downfall.

The technical virtuosity of Italian violinist and composer Nicolò Paganini (1782 - 1840) became legendary.\textsuperscript{26-29} He manipulated his bow exclusively with forearm and wrist, holding the upper arm close to his body. He displayed a phenomenal command of the fingerboard and could move from the first to the highest position at lightning speed either on one string or in four-octave arpeggios, the highest speed recorded being 1 008 notes per minute. Hyperlaxity of his fingers, hands and

<table>
<thead>
<tr>
<th>Table I. Peripartum pelvic pain score (PPPS)</th>
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<tbody>
<tr>
<td>ASLR (maximum 6)</td>
</tr>
<tr>
<td>No weakness (0)</td>
</tr>
<tr>
<td>Subjective weakness (1)</td>
</tr>
<tr>
<td>Objective weakness (2)</td>
</tr>
<tr>
<td>Impossible (3)</td>
</tr>
<tr>
<td>FFT (maximum 3)</td>
</tr>
<tr>
<td>No pain (0)</td>
</tr>
<tr>
<td>Subjective pain (1)</td>
</tr>
<tr>
<td>Objective pain (2)</td>
</tr>
<tr>
<td>Severe pain (3)</td>
</tr>
<tr>
<td>ADD (maximum 6)</td>
</tr>
<tr>
<td>&gt; 150 N (0)</td>
</tr>
<tr>
<td>101 - 150 N (1)</td>
</tr>
<tr>
<td>51 - 100 N (2)</td>
</tr>
<tr>
<td>0 - 50 N (4)</td>
</tr>
</tbody>
</table>

ASLR = active straight leg raising; FFT = femoral flexion test; ADD = adductor strength test; N = Newton.
arms was noted as early as 1831. Early degeneration of shoulder and hip joints caused an aberrant gait. On the stage he looked diabolical. These descriptions are suggestive of the benign joint hypermobility syndrome (BJHS) or Ehlers-Danlos syndrome type III, the benign hypermobile type. Pagani’s advantage of HM, enabling him to play his own compositions, turned into a painful debilitating disease.

**Definition of HM**

Joint HM may be defined as an ‘abnormally increased mobility of small and larger joints beyond their limits of physiological movements, taking in consideration age, sex and ethnic background’.

The maximum range of movement of which a joint is capable is determined by the tightness of restraining ligaments. The primary cause of HM is therefore ligamentous laxity, determined by fibrous protein genes. Joint laxity is maximal at birth, declining rapidly during childhood, less rapidly during the teens, and more slowly during adult life; females are generally 1.5 to 3 times more hypermobile than males.

HM is common in young people, with an incidence of about 5% in a healthy population. Ethnic differences have been described.

HM may represent the upper end of a Gaussian distribution, a *forme fruste* of a heritable disorder of connective tissue (HDCT) such as Marfan’s syndrome, osteogenesis imperfecta and Ehlers-Danlos syndrome, or may be part of other syndromes such as trisomy 21 and congenital myotonia.

**Measurement of HM**

Beighton score criteria are set out in Table II. Each hypermobile joint scores 1 point, the maximum being 9 points. A subject with 5 or more points is considered to be hypermobile. From 1973 till 1998 HM was defined as a Beighton score of > 4/9; from 1998 onwards it has been revised to > 5/9.

**Musicians and HM**

HM may be considered an asset for the aspirant musician because laxity in the fingers, hands and arms will enable easy command of the fingerboard in string instruments or require small movements when playing wind instruments. Larsson et al. found that 63% of prospective flautists, 49% of violinists and 44% of pianists had HM, with percussionists having a prevalence of only 29%. HM was advantageous in those subjects who needed to make rapid, repetitive small movements. Musculoskeletal complaints in hands and arms were reported by 45 - 65% of all musicians, while 77% of those who played string or wind instruments complained of cramps, pain, fatigue and weakness in fingers and arms. Back pain due to ‘loose back syndrome’ was particularly severe in hypermobile female musicians. Premature ‘wear and tear’ caused the joint complaints.

**Table II. Criteria for hypermobility according to Beighton score**

<table>
<thead>
<tr>
<th>Criterion</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hyperextension knee &gt; 10º</td>
<td>2</td>
</tr>
<tr>
<td>Hyperextension elbow &gt; 10º</td>
<td>2</td>
</tr>
<tr>
<td>Passive apposition of thumb to flexor aspect of forearm</td>
<td>2</td>
</tr>
<tr>
<td>Passive hyperextension of 5th metacarpophalangeal joint &gt; 90º</td>
<td>2</td>
</tr>
<tr>
<td>Forward flexion of trunk, with knees straight, so that palms of hand rest easily on floor</td>
<td>1</td>
</tr>
</tbody>
</table>

The benefit of HM with regard to increased ability to play a musical instrument often has to be paid for by early development of degenerative diseases.

**Ballet dancers and HM**

The advantage of HM also holds true for admission to a ballet academy: ballet pupils had a 30% prevalence of HM versus 10% in a control group. Another study found a HM rate of only 9.5%, while our research group found a rate of 35%. Ballet dancers with HM also show premature degenerative changes, especially in the knees.

**Sport and HM**

No published reports on this subject could be found. There is a subjective impression that HM is more frequently seen in horsewomen, athletes and gymnasts (personal observation).

**Ehlers-Danlos syndrome (EDS)**

The cardinal signs of EDS are hyperextensible skin, dystrophic scarring, easy bruising and joint hypermobility. The clinical spectrum of the 10 types of EDS is very variable and the types may overlap: from a mild type with hyperextensible skin and joint HM (EDS II, mitis), via an intermediate type with skin fragility and more obvious HM (EDS III, 'benign' hypermobile type) to the vascular EDS IV type with life-threatening complications such as rupture of vessels, the uterus during delivery, the bowels and spleen. The mode of inheritance is mainly autosomal dominant.

**Pregnancy and EDS type III**

Most authors are of the opinion that common HM is a *forme fruste* of EDS type III. The combination of EDS III and pregnancy has manifold adverse effects. This is readily understood if one realises that an abnormal type or content of collagen type V is the culprit, and that this is present in basal membranes, skin, placenta, muscles, ligaments and uterus.
Maternal outcome

Pelvic pains and instability and/or a diagnosis of PPPP were reported in 26% of pregnant women with EDS as opposed to 9% of unaffected women. Patients with EDS III are most likely to develop laxity (28%); in this group all symptoms of PPPP are more obvious, more luxations occur, and more patients are wheelchair-bound. Caesarean section (CS) should be performed for obstetric reasons and severe symphysiodynia, as the lax symphysis can easily be damaged during vaginal delivery. General anaesthesia is recommended. Closure of the abdominal wall should be as atraumatic as possible and sutures should stay in situ for at least 10 days. Instrumental delivery, especially forceps extraction, should be avoided because of an increased chance of perineal rupture.

Perinatal outcome

Women with EDS III carry a 50% chance of having an affected infant: symptoms in the infant are hypotonia at birth, hernias, luxations and hip dysplasia/dislocation. HM is common in newborns with congenital hip dislocation (30% in girls and 70% in boys). Premature rupture of the membranes also caused an increased rate of premature delivery (25% v. 8% in a normal population).

Benign joint hypermobility syndrome

When HM becomes symptomatic it is called ‘benign joint hypermobility syndrome’ (BJHS). The definition of BJHS is ‘occurrence of musculoskeletal symptoms in hypermobile subjects in the absence of systematic rheumatologic disease’. It has been established that the HM type of EDS (EDS type III) and BJHS are the same.

One may consider this syndrome as late sequelae in HM individuals, such as musicians, ballet dancers and pregnant women. Many older women with BJHS had their first musculoskeletal problems, especially affecting the back, during their first pregnancy. Low back pain during pregnancy seems to be caused by lack of stability due to the increased production of Rlx, which relaxes the pelvic girdle, facilitating vaginal delivery. Most symptoms in BJHS are functional, such as scoliosis, because if the patient bends forward the scoliosis will disappear, as is easily demonstrated in young HM subjects.

Palaeopathological observations of BJHS

Paganini’s BJHS has already been discussed. Studying paintings may reveal signs of BJHS. The ‘Three Graces’ (Fig. 1), painted by Peter-Paul Rubens (1577 - 1640), depicts his second wife Hélène Fourment and her two sisters (Prado, Madrid). The middle Grace has scoliosis with a positive Trendelenburg sign. The Grace on the left shows hyperextension of the distal interphalangeal and metacarpophalangeal joints of the 4th and 5th fingers, and a flat foot. All three Graces have hyperlordosis. These clinical signs are suggestive of BHJS. One would not expect physical abnormalities in the three sensual Graces, but it proves that the artist Rubens (fecit 1638 - 40) was a keen observer long before doctors were.

Results and discussion

Non-pregnant women

The incidences of HM in non-pregnant women are given in Table III. Black African women showed more HM than Caucasians, while ballet pupils showed the highest percentage, confirming other data. HM in the study in schoolgirls was more pronounced in females than in males, confirming data in other studies. Indians in Cape Town had more joint laxity than the Xhosa, who in turn showed more laxity than South Africans of European descent. Although age and pregnancy are important factors resulting in differences in HM, ethnic differences seem to play the most important role. HM in individual joints decreased significantly with ageing, but the overall HM did not diminish significantly.

Pregnant women

The first cross-sectional study in 378 pregnant Caucasian women in the Netherlands revealed that HM was present in 55% of those with complaints of PPPP, while the prevalence in women without complaints...
was only 11.6% (Table IV, total prevalence 15.6%)\(^{30}\). This figure corroborated the hypothesis that pre-existent HM might be the main cause of PFP. However, in studies of South African and Saudi women no correlation was found between HM and PFP. Moreover, severe PFP was virtually absent in both study groups, suggesting that only Western populations suffered from PFP\(^{30,31}\).

The prevalence of HM in pregnant women in the Cape Province was surprisingly low (4.9%\(^{20}\)), while Saudi women had a prevalence comparable to that of Tanzanian women (Table IV). The reason for the large differences between the pregnant women in Tanzania\(^{36}\) and Botswana\(^{35}\) is not clear: the measuring technique was the same, and it may point to different populations.

Low back pain in pregnancy occurred in 38% of the South African\(^{25}\) and 58% of the Saudi women studied,\(^{35}\) but objective measurement using the FFT score gave figures of only 9% and 21% respectively. This proves once again that the FFT test has a low sensitivity, at least in pregnant women.\(^{25}\)

In the South African group symphysiodynia was reported in 7% of cases and the ASLR and ADD tests both had a 6% rate of positive results.\(^{35}\) However, the Saudi study showed a prevalence of 43%, with the ASLR test positive in 23% of cases and the ADD test in 87%\(^{35}\). This group of patients is therefore quite different from the South African group. An explanation could be subclinical hypovitaminosis due to limited exposure to sunlight with resulting proximal myopathy,\(^{35}\) or the high frequency of obesity in the Saudi subjects.

### Table III. Beighton scores in non-pregnant nulliparous women

<table>
<thead>
<tr>
<th>Country (population)</th>
<th>Reference</th>
<th>Age (yrs)</th>
<th>N</th>
<th>N</th>
<th>Beighton ≥ 5/9</th>
</tr>
</thead>
<tbody>
<tr>
<td>Netherlands (Caucasian schoolgirls)</td>
<td>[34]</td>
<td>4-17</td>
<td>44</td>
<td>44</td>
<td>9.8</td>
</tr>
<tr>
<td>Netherlands (Caucasian ballet pupils)</td>
<td>[47]</td>
<td>10-18</td>
<td>53</td>
<td>39.2</td>
<td></td>
</tr>
<tr>
<td>Tanzania (black African schoolgirls and trainees)</td>
<td>[36]</td>
<td>9-36</td>
<td>131</td>
<td>18.6</td>
<td></td>
</tr>
</tbody>
</table>

### Table IV. Beighton scores in pregnant women (cross-sectional studies)

<table>
<thead>
<tr>
<th>Country (population)</th>
<th>Reference</th>
<th>Beighton ≥ 5/9</th>
</tr>
</thead>
<tbody>
<tr>
<td>Netherlands (Caucasians)</td>
<td>[67]</td>
<td>378 59 15.6</td>
</tr>
<tr>
<td>Tanzania (African)</td>
<td>[69]</td>
<td>70 7 10.0</td>
</tr>
<tr>
<td>Botswana (Botswana)</td>
<td>[70]</td>
<td>75 14 18.7</td>
</tr>
<tr>
<td>South Africa (coloured, Cape)</td>
<td>[35]</td>
<td>509 25 4.9</td>
</tr>
<tr>
<td>Saudi Arabia (Arabs)</td>
<td>[68]</td>
<td>202 24 11.9</td>
</tr>
</tbody>
</table>

### Conclusions

1. The hypothesis that women with HM have a tendency to develop PFP during pregnancy could be proved only in the Dutch group. This indicates that populations and their specific ailments are very different.

2. HM is a forme fruste of EDS type III or BJHS.

3. It appears that the advantages of HM in musicians, ballet dancers and pregnant women (‘easy delivery’) may result in severe disadvantages, such as lifelong BJHS.

4. Pregnancy is often a trigger for development of back pain and BJHS.

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\(^{19}\) Chamberlain WE. The symphysial pain in the Roentgen examination of the sacro-iliac joint. *Am J Roentgenol Rad Ther* 1930; 24: 621-626.


\(^{23}\) Van Meeteren J, Mens JMA, Bem J. Ontstekings- naar betrouwbaarheid van het meten van de spiegelhechting van de adductoren bij gemiddelde vrouwen met een daggewrichtsdynamometer (Studie van de reliabiliteit van meting de kracht van de adductoren in gezonde vrouwen met een daggewrichtsdynamometer). Ned Tijdschr Geneeskd 1996; 139: 1573.