Pili bifurcati

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Summary

"Pili bifurcati" is an uncommon hair shaft dysplasia that is often confused with "pili gemini". This is the reason why both hair dysplasias will be dealt with together. The two characteristics that define pili bifurcati are:
1. Each bifurcation produces two separate parallel branches which fuse again to form a single shaft.
2. Each ramus of the successive bifurcations is vested with its own cuticle. When the same papilla changes its shape repeatedly during the anagen phase, it can produce hair shafts with bifurcations at irregular intervals: pili multibifurcati. And, as it is also possible for one of the new papilla to split again in two, the hair shaft may be doubly bifurcated: pili bi-bifurcati.

The term "pili gemini" is used to define a kinetic papilla that during the anagen phase splits at the upper end from single to double-tipped and consequently the same follicular matrix produces two different-sized hair shafts having separate cuticles that emerge through a single pilary canal. Pili gemini maintains the double tipped papilla and consequently the hair shaft does not fuse again. Papillary tips that divide in several tips will produce several hair shafts, that characteristically do not fuse again, might be named "pili multigemini". Patients with pili bifurcati may show diffuse alopecia. The process leading to pili bifurcati is uncommon and occurs in normal hair, in pili canaliculi, monilethrix as well as in the mosaic trisomy 8 syndrome. In the trichogram a "telogen effluvium" must be observed, associated with a high percentage of dystrophic anagen hairs. The calibre of the hair shaft is generally reduced in the 80% and hypopigmentary changes appeared in the 70%. When a "deficiency of good quality protein" is responsible for these dysplasia proteins of good quality in the diet must be administrated.

Keywords: hair shaft dysplasia, pili gemini, pili multigemini, pili multibifurcati, pili bi-bifurcati, anagen phase, cuticle

This review paper aims mainly at clarifying the differences between "pili gemini" and "pili bifurcati". Indeed, while Hurwitz claimed that "pili gemini" and "pili bifurcati" are different disorders which should not be confused (1), others (2,3) believe that "pili bifurcati" represents a restricted form of pili gemini, with a common pathogenesis though. This is the reason both hair shaft dysplasias will be described together. In 1998, Camacho et al. proposed a new classification (4).

Disease characteristics

Pili multigemini
"Pili multigemini" is characterized by groups of hairs of different sizes with their own cuticle.
These hairs grow from the same follicular matrix and emerge through a single pilary canal. The name was proposed in 1951 by Pinkus (5) who indicated that two to eight shafts may grow from a single follicular matrix and papilla, whose upper end is split into tips corresponding one-to-one with each hair shaft.

**Pili gemini**

When there is only one bifurcation of the papilla or matrix, two hair shafts emerge through the pilary canal; then it must be named "pili gemini".

**Pili bifurcati**

"Pili bifurcati" was described by Weary et al. in 1973 (9). This uncommon developmental defect of the hair growth is characterized by intermittent bifurcation of the whole hair shaft at irregular intervals (10). Each bifurcation produces two separate parallel branches which fuse again to form a single shaft. Each branch of the successive bifurcations has its own cuticle, which completely surrounds it (11). The anomaly appears to be transitory, with only a small percentage of hair exhibiting the bifurcation (1). The patient with this type of anomaly may show diffuse alopecia, more evident in some areas, similar to trichotillomania or pili torti (2,11).

**Pili multibifurcati**

If the matrix-papilla changes its shape several times during the anagen phase, it produces hairs with bifurcations at irregular intervals. Like in the previous situation, each branch of the successive bifurcations has its own cuticle, which surrounds it completely (figures 1d, 5).

**Pili bi-bifurcati**

It is even possible that the same matrix-papilla which was already divided, might subdivide again, leading to a bifurcation of the already bifurcated branch. Each new branch has its own cuticle, which surrounds it completely, and like in the single bifurcation the branches will be of different size or diameter. Finally the branches of the second bifurcation fuse again, and afterwards the first bifurcated branches fuse again. This disorder can be named "pili bi-bifurcati" (figures 1c, 4).

**Pathogenesis**

**Pili gemini and pili multigemini**

The pathogenesis of this dysplasia is as follows. During the anagen phase, a kinetic dermal papilla changes its form from single-tipped to double-tipped producing two hair shafts that separately emerge through the same pilary canal (figures 1a, 2). This process is very common, and can be observed both in other dysplasias and in normal hair. Less frequently, the papilla splits into four or eight tips producing four to eight subpapillae which will produce 4 to 8 hair shafts that emerge separately from the pilary canal, leading to pili multigemini.

**Pili bifurcati**

During the anagen phase, the matrix-papilla changes its form from single-tipped to double-tipped, then back to single-tipped again; consequently, a single hair shaft is produced, then a bifurcated hair shaft, and then a single hair shaft again. Each branch has its own cuticle and they are of different diameter. This dysplasia should be named "pili bifurcati" (figures 1b, 3).

**Pili bi-bifurcati**

Although an autosomal recessive inheritance was proposed, this mode of transmission needs to be confirmed (16).

**Pseudo-pili bifurcati vs central trichoptilosis**

This form is not a true pili bifurcati since the split sections are not surrounded by the cuticle. It would be called "central trichoptilosis". Nevertheless, it has been shown in several trichological studies, generally associated with other hair shaft dysplasias (17,18), and it was named "pseudopili bifurcati". In my department, all dermatologists have seen this disorder in several dysplasias, and also in trichonodosis in normal hair with knots produced by mechanical or physical forces (figures 1e, 6) (19). Since it is absolutely different from "pili bifurcati" because it is produced as a consequence of a trauma of the hair shaft, resulting in splitting with two parts that are, when taken separately, never surrounded by a complete cuticle, and the cortex and medulla may be broken, and, in addition there are not changes in the matrix-papilla, the names "pseudo pili bifurcati", "acquired pili bifurcati" or "iatrogenic pili bifurcati", must be avoided and changed by "central trichoptilosis".

**The various designations**

Although we admitted that the bifurcations of pili bifurcati are a consequence of splits of the follicular germs, and consequently that "pili bifurcati" would be the result of the "pili gemini", this "terminology" must not be applied to all the forms of pili bifurcati. In addition, normally, the diagnosis is by made by trichogram, in other words, on the hair shaft or "pili" and no by biopsy or on the matrix-papilla (germs). But we must use both terms to define different disorders.

**Pili gemini**

Pili gemini should be used to define situations in which the matrix-papilla is split producing two different branches (pili gemini) or four to eight different branches (pili multigemini) that not fuse again since they will be separate and independent permanently.

**Pili bifurcati**

Pili bifurcati would be used when the branches of different sizes fuse again, once (pili bifurcati), several times (pili multibifurcati), and once again on previously bifurcated branch (pili bi-bifurcati),
independently that the kinetic matrix-papilla must be transformed again from two into one papilla.

**Central trichoptilosis**
In "central trichoptilosis", neither splitting of the matrix and papilla nor true bifurcations are observed. Hence, this acquired condition should be clearly separated from "pili gemini" and "pili bifurcati".

In accordance with these concepts, Camacho *et al.* proposed a new classification in 1998 (4)(table 1).

**Table 1. The classification by Camacho *et al.* of Pili bifurcati (1998)**

<table>
<thead>
<tr>
<th>Disease name</th>
<th>Hair fuse again</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pili gemini</td>
<td>No</td>
<td>Change of one papilla into two which are maintained</td>
</tr>
<tr>
<td>Pili multigemini</td>
<td>No</td>
<td>Change of one papilla into four to eight or sixteen subpapillae which are maintained</td>
</tr>
<tr>
<td>Pili bifurcati</td>
<td>Yes</td>
<td>Change of one papilla into two and posterior fusion</td>
</tr>
<tr>
<td>Pili multibifurcati</td>
<td>Yes</td>
<td>Intermittent changes of one papilla into two and from two into one</td>
</tr>
<tr>
<td>Pili bifurcati</td>
<td>Yes</td>
<td>Change of one papilla into two and posterior subdivision of one of these into another two</td>
</tr>
</tbody>
</table>

**Occurrence in other syndromes**

**Pili multigemini**
Pili multigemini has been reported in cleidocranial dysostosis (6) and in the trichorhinophalangeal syndrome (7).

**Pili bifurcati**
The process leading to pili bifurcati is uncommon, and occurs in normal hair and in pili canaliculi and monilethrix. It was also described in association with the mosaic trisomy 8 syndrome (12).

**Frequency**

**Pili gemini and pili multigemini**
"Pili multigemini" is rare in daily practice, and when clinically several hair shafts are found emerging from a single pilary canal, the differential diagnosis of "trichostasis spinulosa" must be considered (8).

**Pili bifurcati**
This dysplasia is very uncommon and has been only diagnosed four times in my department in cases secondary protein deficiency (13). In these cases of "high-quality protein deficiency" (14), after a telogen effluvium, the follicular matrix-papilla has to produce a new hair shaft. During this new anagen phase, the kinetic matrix-papilla would divide into two, to produce a bifurcated hair. This would later, when deprived of good quality protein again transform into a single matrix-papilla. Afterwards, on recovery, this would again subdivide, and so on, thus showing intermittent bifurcations.

**Pili bi-bifurcati**
It is extremely uncommon, and was described in a four-year-old boy with alopecia in a large area of his occipital region (15).

**Diagnostic methods**
The pull hair sign is positive in any cases, especially in cases of secondary protein deficiency but the Sabouraud and Jacquet signs are habitually negative.

In the trichogram a "telogen effluvium" must be observed, associated with a high percentage of dystrophic anagen hairs. The calibre of the hair shaft is generally reduced in the 80% and hypopigmentary changes appeared in the 70%. The biopsy can be useful in "pili gemini" or "pili multigemini" but in "pili bifurcati" and its different forms, the only clear exploration is the "hair shaft study" when, of course the hair shaft must be obtained by cutting. The scanning electron microscopy must be used but it would be lucky to observe these dysplasias.

**Management**
When a "deficiency of good quality protein" is responsible for these dysplasia proteins of good quality in the diet must be administrated. Occasionally, iron deficiency could be demonstrated and in these cases treatment with iron must be started at least during six months. Patients must be always told to avoid mechanical procedures for care of their hairs.

**References**