



## Pilonidal sinus disease - Etiological factors, pathogenesis and clinical features

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### ABSTRACT

'Pilonidal sinus' disease, which is most commonly seen in reproductive populations, such as young adults - mostly in males who are in their twenties - is actually a controversial disease in that there is no consensus on its many facets. It is sometimes seen as an infected abscess draining from an opening or a lesion extending to the perineum. It may also present as a draining fistula opening to skin. In terms of etiological factors, various theories (main theories being congenital and acquired) have been established since it was first described, no universal understanding achieved. A long and significant post-operative care period with different lengths of recovery depending on the type of operation are quite prevalent with regards to recurrence and complication status. In order to prevent recurrence and improve the quality of life, etiological and predisposing factors as well as clinical features of sacrococcygeal pilonidal disease should be well known, a detailed differential diagnosis should be made, and a suitable and timely intervention should be performed. It was aimed here to explain the etiological factors, pathogenesis and clinical features of the disease that may present with various clinical symptoms.

**Key words:** Pilonidal sinus disease, clinic, etiology, pathogenesis

### Introduction

'Pilonidal sinus' disease, most commonly seen in reproductive populations like young adults - mostly in males who are in their twenties - is actually a rather controversial disease in that there is still no consensus on its treatment, probability of its recurrence is high, and its care takes a long time and is costly. It is mostly an acquired, chronic and inflammatory condition affecting the sacrococcygeal region in most cases. It is sometimes seen as an infected abscess draining from an opening or a lesion extending to the perineum. As well, it may also be observed as a draining fistula open-

ing to skin [1]. In this article, the aim was to explain the etiological factors, pathogenesis and clinical features of the disease that may present with various clinical symptoms and impairs life quality and satisfaction [2].

### Etiology

Although various theories have been established since it was first described, no consensus has been reached.

According to the supporters of congenital theory, pilonidal sinus disease occurs because of residual epithelium in the spinal canal and skin or when hair follicles enter to interspace formed by incomplete fusion

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occurring as a result of a defective union of the skin layer during the early embryonic period. On this basis, from the time of Fere (1878) to Stone (1951) and even until today, a number of authors have established four different theories: medullary canal theory, dermal inclusion theory, beak glands theory and sex gland theory [3]. These theories, recently having just few supporters, have never been proven and failed to explain pilonidal diseases located in the regions other than the sacrococcygeal area [4,5]. These authors have been known to misdiagnose certain diseases, such as medullary canal fistulas and hydroadenitis, important in the differential diagnosis of pilonidal disease. In addition, successive clustering of hairs with the same length, thickness, colour and position could not be explained by this theory [6, 7]. Cubic epithelial lining of the inner wall of congenital pilonidal disease undermines this theory [8]. The main characteristic of pilonidal sinus is squamous epithelium lining its inner wall.

According to the supporters of acquired theory, pilonidal disease occurs as a result of inflammation that results from foreign body reaction occurring secondary to the entry of hairs in the subdermal area after trauma. Supporters of this theory also suggest various other theories to explain the occurrence of this mechanism. According to Bascom, hair follicles become infected under the effect of sex steroids when they are filled with keratin during puberty, this infection forming an abscess tract. Hairs in this tract pierce the skin while they are still attached to it and are pulled into deeper regions under the vacuuming effect of gluteal muscles to form a sinus [7]. The vacuuming effect is mostly observed in the sitting position. For this reason, it is suggested that this disease is more common in people that sit for long periods of time. However, this disease is not very common in patients confined to bed and elderly people who spend their time mostly sitting during the day. Karydakos suggests that hairs move towards the follicle and enter into deeper regions like a drill. The first entry occurs through a weak part of the skin and other hairs come through after [9]. This theory is validated by the fact that the disease occurs in interdigital areas, the umbilicus, clitoris and anal canal, and it recurs even after radical excisions.

### Pathogenesis

Pilonidal sinus is a chronic inflammatory disease that develops between the sacrococcygeal joint and coccyx. It is mainly composed of 2 canals - one canal that begins from the sinus orifice that opens to skin and moves upwards subcutaneously for 2-5 cm, called the primary canal. Canals which connect the canal located posterior to primary canal with the skin are called secondary canals. Resultant abscesses initially form in secondary canals and then primary canals. The primary canal is lined by squamous epithelium, while secondary canals are lined by granulation tissue. Hairs freely located in the primary canal and secondary canals contain hair follicles, leukocytes, hemosiderin-containing macrophages but not sweat from sebaceous glands [10]. In a typical case, hairs gush outwards from the sinus opening, and unlike the anal fistula, these sinuses usually move upwards and towards the sacrum, mostly unable to reach to the bone and end as a blind and enlarged cavity. This cavity is macroscopically seen as a cystic structure. Hairs may appear as they gush outwards from the sinus or may be first seen in the wound after it is opened. When these hairs are examined, they are seen as structures with thin edges as the hairs shed. However, in a study by Kooistra, only half of the cases were found to have hairs in the wound [11, 12].

Bascom suggested that the origin of pilonidal sinus disease was pits could not be present without them. They might best be seen in the midline when the gluteus is opened to both sides. Pits with diameters reaching up to 1-3 mm are formed by hair follicles attached to the skin with the help of the power of the vacuum effect. The weakest point of the skin is pierced and the abscess process begins. Bascom also put forth that keratin is important in the pathogenesis of pilonidal sinus disease, such that keratin shed from the stratum corneum enters into the pits with the effect of pressure and compression, contributing to the formation of an abscess and inflammation by anaerobic bacteria. The analogy is akin to keratin acting much like the match making the fire [7, 13].

Further, Bascom proposed that in addition to hair follicles, cut or shed hairs also had roles in pathogenesis. Rather than the type and quality of the hair, the



Figure 1. Asymptomatic form.



Figure 3. Chronic disease form.



Figure 2. Acute abscess form.



Figure 4. Recurrent pilonidal sinus disease.

place where hair grows is important. It does not make any difference whether it is located either above or under the skin, but hairs piercing the skin facilitate the establishment of the infection. Karydakis formulated the idea that hair shedding, piercing ability and affectability of the skin in the natal cleft as an important triad in the pathophysiology of pilonidal sinus disease [14]. For this reason, surgeons want to leave a flat surface in the area, rather than a nook surface to which hairs may attach and accumulate.

Compression and deoxygenation, accepted as another important pathophysiological cause of pilonidal disease. Natural compression and depth of natal groove prevents the drainage of abscess located in deeper regions and above all, they keep healing oxygen away from the area. Oxygen cannot be utilized for oxidative killing, which is an important defence mechanism, and so disease recovery becomes difficult [15].

### Clinical Findings

Pilonidal disease has 3 different clinical presentations:

1. Asymptomatic form
2. Acute abscess form
3. Chronic disease form
  - Recurrent complex pilonidal sinus disease;
  - Chronic pilonidal sinus disease.

**1- Asymptomatic form:** patients have no complaints and are incidentally diagnosed. One or more pits are located at the midline, about 5 cm from anus, are detected (Figure 1). In a study by Eftaiha and Abrasian, the ratio of these patients among all patients with pilonidal sinus was found to be 11% [16].

**2- Acute abscess form:** severe pain, swelling and redness are present in the sacrococcygeal region (Figure 2). Fever and leukocytosis may be present. It is the most common cause of hospital admission [17]. Another study showed that 50% of patients presented



**Figure 5.** Post-operative appearance.

with this form [18]. The indurated area containing abscess gradually grows and accumulated malodorous fluid spontaneously drains or is surgically drained. After this stage, conversion to the chronic form begins. Microbiological examinations of drainage fluids mostly reveal anaerobic proliferation (*Escherichia coli*) [19]. Other common microorganisms are known to be *Proteus*, beta hemolytic *Streptococcus*, *Pseudomonas* and *S. aureus*, respectively. There are also studies that suggests *S. aureus* and *Streptococcus* were more common [20]. In a study by Sondena et al., the growing microorganisms in abscess cultures were anaerobes (mainly *E. Coli*) in 77%, aerobes in 4% and mixed microorganisms in 17% of the patients [21].

**3- Chronic Disease Form:** Chronic Pilonidal Sinus Disease: named after the period of abscess drainage. Intermittent drainage and pain are present (Figure 3). Mild tenderness and induration is present upon palpation. There may be one or more than one sinus orifices on which hairs may grow. Skin may have shrinkage and scarring secondary to drainage.

**Chronic Recurrent Pilonidal Sinus Disease:** recurrent abscess attacks following drainage lead to the formation of independent and irregular orifices and scars. It is the name of this condition. As a result of these acute exacerbations, irregular fistulas sometimes move towards the anal canal and rectum to cause the formation of complete fistulas and the sacrum may even be infiltrated because of extension of sinus tissue. It is a condition which may cause psychological problems that negatively affects one's social life results in a decrease in quality of life (Figure 4).

## Discussion

The literature suggests that malignant degeneration - especially squamous and varicose carcinomas - may occasionally develop on the basis of disease with long-standing drainage and laceration [22]. It is recommended that wide excision should be performed for chronic, non-healing, ulcerated and rapidly growing lesions and involved inguinal lymph nodes should be biopsied. In the cases with metastatic lymph node involvement, recurrence rate is 50%, 5 year survival is around 50% and prognosis is poor. Chemoradiotherapy may be added to the treatment regimen in these cases [23].

Pilonidal sinus disease should be differentiated from those diseases which involve the sacrococcygeal region [24];

- pruritis ani;
- carbuncle-furuncle, pyoderma gangrenosum;
- periproctitis and perianal abscess and fistulas;
- tuberculosis granuloma, actinomycosis
- osteomyelitis drained by sinuses,
- syphilis,
- spina bifida, medullary canal fistulas
- hidradenitis suppurativa, postanal dermoid cysts.

It is very important for prognosis to make a differential diagnosis among these aforementioned diseases that have different diagnostic and therapeutic protocols (Figure 5).

Long and important post-operative care periods and different lengths of recovery depending on the type of the operation are quite discouraging in terms of recurrence and complication status. In order to prevent recurrences and improve the quality of life, etiological and predisposing factors, and also clinical features of sacrococcygeal pilonidal disease, should be made well known, a detailed differential diagnosis should be made, and a suitable and timely intervention should be performed.

## Conflict of interest statement

The authors have no conflicts of interest to declare.

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