INTRODUCTION

Pilonidal disease (PD), or the epithelial coccygeal passage (ECP) is defined as a thin epithelial tube, located in the subcutaneous tissue of the intergluteal cleft, at the level of the coccyx, or less frequently at the level of the sacrum. The disease is characterized by recurrent, or chronic inflammation. Often ECP is 2-5 cm in length and ends blindly in the soft tissue at the top of coccyx. On the skin it can present as a single, or less frequently as multiple spot-like, round openings in the intergluteal cleft. The openings are frequently located within 0.3 – 1 cm from one another. The primary openings often have tufts of hair growing from them. Additionally, PD can manifest as secondary, abscess-like fistulas.

The first description of PD appears in the works of A. W. Anderson in a letter to Boston Medical and Surgical Journal, titled “Hair Extracted from an Ulcer”. Anderson described a fistula that contained hair, which he found near the coccyx of his patient [1]. Later, in 1854, J. M. Warren added to the works of Anderson, by describing PD in greater detail, providing guidelines for treatment, as well as proposing the first hypothesis for the development of PD. Warren thought that the etiology involved atypical hair growth (from superficial to deep, instead of the normal deep to superficial) in the intergluteal cleft. The openings are frequently located within 0.3 – 1 cm from one another. The primary openings often have tufts of hair growing from them. Additionally, PD can manifest as secondary, abscess-like fistulas.

The selection of literature was based on such concepts as: etiopathogenesis, historical principles of treatment, methods of surgical and non-surgical intervention. Data from metanalysis publications and randomized clinical trials pertaining to the treatment of the pilonidal sinus at various stages of its formation was used, as well.

KEY WORDS: pilonidal disease, analysis, literary sources
ETIOLOGY AND PATHOGENESIS

Often, PD is asymptomatic for long periods of time, as it doesn’t cause, the child or his/her parents any discomfort. Still some parents choose to consult a surgeon with concerns over apparent openings near their child’s coccyx, or sacrum. PD is also often discovered during routine health maintenance procedures. According to some authors, PD should not be considered a disease due to its asymptomatic nature. Rather, according to these authors, PD should be considered a disease from the time of its first clinical manifestations, which present in the form of an abscess in the sacrococcygeal region.

Discussions over the etiology pathogenesis of PD continue since its first description. It’s worth noting that researchers residing in post-Soviet countries exhibit cardinally different views of PD than their Western counterparts. This tendency is reflected in the different approaches to treatment of patients with PD.

The existing theories of PD etiology and pathogenesis can be divided into several categories: I – historical theories, II – the neurogenic theory, III – the ectodermal theory, IV – the theory of acquired PD.

The first category has strictly historical significance, as it’s based on the propositions of physicians living in 18th – early 20th century, which were predominantly not based on clinical findings. For example, J. M. Warren in 1854 explained the appearance of fistulas in the coccygeal region by pathological behavior of hair follicles in the same region.

The neurogenic theory has found its reflections in the works of F. G. Mallory (1892), who found neural tube remnants lined with cylindrical and cuboidal epithelium in embryos during 3-6 months of gestation. M. Cage (1939) has found in his research a 2-chambered cyst in the coccygeal region. The cysts were connected by a passage, which he proposed exhibited neurogenic etiology. In 1887 E. Tournex and G.J Hermann have proposed that the epithelial coccygeal passage develops from rudimentary spinal cord remnants. During embryogenesis the cranial end of the spinal cord (which forms the brain), and the caudal end (which attaches to the skin in the intergluteal fold) becomes fixed. During the 3rd month of gestation, uneven growth of the spine and the spinal cord is noted, as a result of which, the latter eventually becomes thinner. During further development, normally, the thinned region of the spinal cord (filum terminale) partially involutes, and partially remains in the form of a trough lined by cylindrical epithelium. This trough will normally disappear by the end of the 6th month of gestation. In the case of failure of this trough to involute PD may arise.

Various theories of congenital PD are often grouped together under “ectodermal origin”. The beginnings of the theories of ectodermal origin can be traced back to the works of O. Lannelongue (1882), who proposed that PD arises as a result of ectodermal invagination into the intergluteal fold. This invagination, Lannelongue thought, occurs due to disproportional development of subcutaneous fat tissue in the coccygeal region, as a result of abnormal skin attachment within the region. Independently, C. Gusenbauer (1893) and E. Crone (1917) have discovered that pilonidal cysts exhibit tissues of dermal origin, complete with hair follicles and sweat glands.

H.B. Stone (1931) connected the development of PD to changes observed in puberty. Stone made his observations in the uropygial glands, which are secondary sex organs in avian species [9, 10].

The findings of L. Tail (1924) are considered to be the most well founded, and well-studied. Tail proposed that the invagination of ectoderm towards the floor of the intergluteal cleft is connected to the reduction of coccygeal vertebrae (theory of tail’s ligament). This theory is based on the particular pattern of development seen in the caudal end of the spinal column. During the 5th-6th week of gestation the coccyx is composed of 9 vertebrae, which during further development will reduce to only 5 vertebrae. If this reduction does not occur, pilonidal cysts may form.

In modern literature, the theory of acquired PD has gained popularity. This theory encompasses several different proposed mechanisms of PD development. R. M Hodges (1880) is considered to be the first to propose the acquired etiology of PD. Hodges stated that the PD arises as a result of entrapment of hair follicles within the coccygeal passage. He further noted that these trapped follicles originate from neighboring skin regions, rather than from within the coccygeal passage itself. R. M. Hodges has also stated that the presence of asymptomatic coccygeal passages in itself should be considered a pathology, rather the entrapment of hair follicles within them, and subsequent inflammation of the passages is what defines Pilonidal Disease. In the later years, studies of the anatomical and physiological features of the sacrococcygeal region have been conducted, in an effort to explain how these features may effect the development of PD. In 1934 M. Burgdorf has turned his attention towards connective tissue adhesions between the sacral bone and the skin. He also noted that excess development of subcutaneous fat in this region leads to the development of traction diverticula, in which inflammation may arise as a result of accumulation of the various metabolic products of the skin.

In 1952 W.P Kleitsch and L. Cherry connected PD with such morphological features as a narrow pelvis, and a deep intergluteal fold [11]. These factors, according to the authors, promote friction, excess sweating and maceration within the intergluteal fold, which lead to the formation of a “weak spot”, into which microbes and hair may penetrate.

In 1954 O.N. Davage proposed that PD is of inflammatory origin, stating that hair may be a secondary factor in the pathogenesis, while inflammation within the intergluteal fold should be considered the primary factor, due to which the epithelial coccygeal passage forms [12]. O.N. Davage considered mechanical injury of the epithelium, namely by use of coarse toilet paper to be one of the key factors in the formation of pilonidal cysts. This notion was later supported by R.M. Hardawa (1958), who found small pieces of toilet paper inside a pilonidal cyst [13].

In 1959 R.A. Raffman connected the development of PD to the penetration of hair into the macerated skin,
however, he proposed that the subsequent epithelization of these macerated regions may be due to the epithelial cells, which have been introduced into the region together with the hairs [14].

A year earlier, D.H. Patey and R.W Scarff (1958) demonstrated that the presence of hairs within the epithelial coccygeal passages isn't simply due to chance, but rather due to the presence of negative pressure within the lumen of the epithelial passage, which in turn can be explained by rhythmic contraction and relaxation of gluteal muscles [15]. According to the researches, this pump-like mechanism may explain the high incidence of PD among American troops (predominantly those serving in the mechanized infantry platoons) during the Second World War. The disease was seen especially often in Jeep drivers. This gave PD one of its many names – “the Jeep Disease” [16].

In 1980 J. Bascom proposed the follicular retention theory of acquired PD, which is widely accepted today among most researchers. The differentiating characteristic of this theory is that the author was the first to propose a direct connection between the formation of pilonidal passages and inflammation of hair follicles, located in the midline of the intergluteal fold floor. The Author describes in detail a “trichogenic-pump” mechanism of secondary formation. Bascom further proposes 6 stages of PD development, characterizing each stage by morphological findings:

Stage I (a dilated hair follicle) – excess accumulation of keratin takes place, often as a result of some precipitating factor (increased production of keratin, as a result of hormonal imbalances during puberty, decreased outflow of keratin as a result of the tight closure of the intergluteal cleft in the sitting position, inflammation as a result of poor personal hygiene, an inflammation and swelling due to injury, or bunt force trauma of sacrococcygeal region)

Stage II (an infected hair follicle, or acute pyogenic folliculitis) – an infection of the follicle occurs, with subsequent spread of the infection to the neighboring subcutaneous fat tissue.

Stage III (an acute pilonidal abscess) – the hair follicle ruptures at its weakest point, more often at its base, due to excess pressure during sitting, or due to multiple force vectors in the standing position.

Stage IV (formation of a primary fistula, or the development of a chronic pilonidal abscess) – the inflammatory exudate drains, while the acute process becomes chronic, preventing the fistula from healing.

Stage V (formation of a primary epithelial passage) – the epithelial passage develops from cells of the basal cell layer beneath the ruptured follicle.

Stage VI (formation of secondary epithelial passages) – propagated by the “trichogenic pump”, which first leads to the accumulation, and subsequently to the penetration of hair from neighboring skin regions into the primary epithelial passage. This process occurs via the “fish hook” mechanism (cutaneous protrusions allow the movement of hairs in only one direction). Concurrently, there is inflammation in the underlying subcutaneous fat tissue, which leads to the formation of a secondary acute abscess, which in turn drains, forming a secondary fistula. This secondary fistula becomes chronically inflamed (secondary chronic pilonidal abscess). In the absence of adequate treatment, eventually the walls of the secondary fistula will undergo epithelization with cells from the surface of the skin [17,18,19,20].

J. Bascom’s theory was supported and further developed by G.E. Karyadaki, who in 1992 published his own research based on the observation and treatment of 6000 patients. He noted that cut hairs, imbedded in a normal tissue can trigger a foreign body response. Furthermore, Karyadaki formulated a theory of PD pathogenesis based on 3 variables: shaved hairs (H), which is acted on by some force (F), which is the result of some secondary factors, such as depth of the intergluteal fold, width of the gluteal fold, and the amount of friction within it. This force is what causes the penetration of shaved hairs into the fistula. The third factor is some sustained trauma of the skin and the soft tissues (V) [24]. Therefore, the formula reads as:

PD = H*F*V^2

A.V. Kulyapin (1989) found by investigating the hair follicles and the sebaceous glands under the microscope, the latter were without changes. Conversely, he has found notable changes in the observed sweat glands, from the same region. Based on these findings, Kulyapin proposed that acute Hidradenitis Suppurativa, which arises secondary to blockage of sweat gland outflow channels by hairs and various other debris may be the cause of PD. It is worth noting that sweat glands in the intergluteal folds are apocrine, and so they begin to function at the beginning of puberty.

METHODS OF TREATMENT

Radical surgical treatment is currently the universally accepted method of treatment of PD. However, even today there isn’t a single universal approach which satisfies both; the physician, and the patient. Therefore, specialists within the surgical and coloproctological fields continue developing new surgical approaches. In these developments, the researchers seek to decrease the recurrence of disease, especially during periods of remote observation. Researchers seek to minimize the amount of postoperative pain, and the frequency of postoperative infection. Finally, they aim to decrease the amount of time for which the patient must remain in the hospital following the operation, likewise, they seek to minimize the recovery time period [21,22].

Overall, all existing surgical treatments of PD can be divided into 4 categories: conservative surgical intervention, open surgical intervention, closed surgical intervention, and minimally invasive surgical intervention.

Acetic acid, Ethanol and Chloroform, as well as other so-called sclerosing agents are used to treat PD non-surgically. Electrocoagulation, and cauterization with acetic acid or nitrous oxide have remained common options until the end of the 20th century [24]. In 1956 A.A. Klaus only used strict measures of hygiene, as well as shaving in the treatment of his PD patients. According to Klaus, he saw...
recovery in 89.3% of all cases [25].

B. Maurice and R. Greenwood were known components of conservative treatment methods. In 1964, they performed injections of 40% and 80% phenol into their patients’ coccygeal passages. [26]. Subsequent randomized trials, which compared the phenol injection method to surgical intervention, have shown that recurrence of disease was higher in the case of the former. However, conservatively treated patients noted less pain, and were able to return to carrying out their normal daily activities sooner, compared to those who were treated surgically [27,28]. Later D. Ahmet [2012] treated 83 PD patients with 80% phenol, and found that on average, the patients were able to return to their normal daily activities in 2-3 days, while there was a 13.3% rate of recurrence [29].

Open surgical approaches to the treatment of PD imply excision of the pilonidal cyst, achievement of hemostasis, and treatment of the cavity with antiseptic agents. The wound remains open, which leads to healing by secondary intention. This technique has been reported to show a low rate of PD recurrence, as well a low rate of infectious complications. This is attributed to the unobstructed drainage of fluid from the wound’s surface. Another advantage of open surgical approaches is the relative simplicity of the techniques, which causes these techniques to be favored in many countries around the world. One notable disadvantage of the open surgical approaches is the relatively long period of time the patient must remain in the hospital, due to the long time required for complete healing of the wound (several months, or more). At the same time, the patients tend to dislike the visual appearance of the scar (which often undergoes keloidal transformation, and ulceration). Patients also tend to experience itching, reduced mobility in the region, and discomfort in the sitting and squatting positions [30].

In order to reduce the duration of patients’ stay in the hospital, and the over-all healing time, a wide variety of modified open approaches have been proposed. These modified techniques imply the suturing of the wound’s edges to its floor, thus reducing the wound’s area, and reducing the duration of time, in which the wound heals by secondary intention. These techniques have been demonstrated to only marginally reduce the time spent in hospital, meanwhile no notable improvements along other parameters have been noted at all. This category of surgical approaches includes the so-called “marsupialization of the pilonidal cyst”, and its many variants, proposed by Obeid, McFee, and others [31].

In 2014 I. Varnalidis et al published their research, in which they compared 3 methods of surgical treatment of PD: the open approach, the marsupialization approach, and the closed approach with complete suturing of the wound along the midline. The researchers have determined that the average amount of time the patients took to recover after the marsupialization method was 27.3 days, with a maximum of 40 days. The closed approach with tight suturing along the midline showed a recovery time of 11.7 days, with a maximum of 15 days. Finally, the patients recovered in 46.4 days, on average (with a maximum of 90 days) after the open surgical approach with healing by secondary intention. The research also noted the rates of PD recurrence, which for the marsupialization method, and the open approach were observed to be 6.35% and 3.45%, respectively. Meanwhile the rate of PD recurrence after the closed approach with tight suturing was a much higher 57.8% [32].

The closed, midline suturing approach falls under the third category of surgical PD treatment. The technique is also relatively wide-spread around the world, mainly because of its simplicity. After the cyst is excised, the wound is sutured with a simple surgical knot along the midline. This approach, and all other closed approaches encourage healing of the wound by primary intention, thus decreasing post-operative recovery time, and decreasing the size of the scar. One notable disadvantage of this technique is a high rate of PD recurrence, and a high rate of infection. This can be attributed to obstructed draining, increased tension along the suture line, and increased friction between tissues in the area [33].

In 2015 Doll D. et al published their analysis based on findings made in the treatment of patients with PD over 20 years. In their analysis, they compared the outcomes of open and closed surgical approached. According to the data, the recurrence rate after 20 years of observation post-surgery was higher in the closed approached with tight suturing, than in the open approach, with rates of 44% and 28%, respectively. Also, according to the gathered data, the researchers have concluded that the though the separation of wound edges during the healing process lengthens recovery time, it has no statistically significant influence on the rate of PD recurrence, at the 5 and 10 years of observation [34].

A Al-Khamis et al (2010) have composed a meta-analysis of PD patient treatment with various open and closed surgical approaches, in which the authors have found that when comparing the two, neither showed any significant advantage over the other. As with other similar studies, when assessing the duration to complete healing, closed approaches have been found to take on average 15 days to heal, while the open approaches took on average 41 days to heal. Conversely, open techniques have shown a much lower rate of PD recurrence (12.5% against 24% observed in closed techniques). No statistically significant difference in rate of post-operative infection was noted between the two. However, a significant advantage of the “lateralization technique”, which is based on suturing laterally from midline, was shown over the tight midline suture method. The lateralization technique demonstrated healing times reduced by 5.4 days, compared to the tight midline suture technique. Frequency of infection was also noted to be reduced by 3.72%, while frequency of recurrence was observed to be reduced by 4.54% [35].

Proponents of the acquired origin theory have proposed a new view on the current methods of surgical intervention, stating the importance of forming the postoperative wound on the buttock, instead of at the floor of the intergluteal
cleft. They argue for this approach based on the assumption that a surgical wound formed at the floor of the cleft is under the influence of many unfavorable forces. Instead, these researchers propose moving the wound by 3-4cm laterally, thus somewhat flattening the intergluteal cleft, and according to them, reducing the possibility of hairs entering the wound and formation of a "residual cavity". The latter can be explained by better alignment of wound edges and the decrease in tension between tissues [42,43].

G.E. Karydakis (1973) first proposed the use of the aforementioned "eccentric" suturing technique, with mobilization of two "flabs" of skin after the pilonidal cyst has been excised. This method has gained popularity for its reported high efficacy. Later, in 1992, the author has provided a convincing base of evidence, grounded on the treatment of over 6500 patients, according to which, the rate of PD recurrence was 1%, rate of complications – 8% and the amount of time spent in hospital was on average 3 days, with a home recovery time of 15 days on average [36].

Subsequently, other researchers have attained date similar to that of G.E Karydakis. Can et al [37], in their research have found frequency of PD recurrence to be 4.6%, while post-operative complications were noted in 8.69% of all cases [37]. According to the data gathered by S. Sozen et al (2010), the rate of recurrence was noted to be at 1.5%, while the rate of complications was noted to be 6% [38]. M. Sakr et al (2003) have reported a 2.4% rate of recurrence, while complications were noted in 10% of patients during their in hospital stay (3 days). Additionally, the Karydakis method was shown to be highly effective in treating obese patients (BMI > 26.5 kg/m²) [39].

The "rhomboidal plastic method" proposed by A. Limberg in 1946 is currently another widespread surgical technique for the treatment of PD. This method is characterized by resecting the pilonidal cyst rhomboidally and covering the surface of the wound with an accordingly shaped piece cutaneous-subcutaneous tissue, resected from the buttock. This placement of 2 adjacent triangular pieces of tissue over the wound provides an adequate redistribution of tension. The main advantage of this technique is in the low rate of recurrence and postoperative complications [23].

In 1929 F. Lahey was first to successfully cover the surgical wound after resecting a pilonidal cyst with a cutaneous-subcutaneous pedunculated graft. However, this approach saw a large number of recurrences, and was a fairly complicated procedure over-all, and therefore never gained widespread popularity [60]. R.H Fishbein (1979) performed a closing of the defect with a cutaneous-subcutaneous-fascial graft, which allowed for a decrease in recurrence to 2.5% over 36 months of postoperative observation [56]. R.S Monroe and F.T. McDermott (1963) used a so-called "Z-like plastic method", the results of which were inconclusive: despite the low rate of recurrence (up to 8.9%), major postoperative complications, such as necrosis of the graft (2.5%), formation of keloids (up to 14%), diminished sensitivity of the soft tissues, and unsightly scars (up to 50%) were noted [57,58,59,60]. R.F Roth et al (1977) proposed a "W-like plastic method", which also never gained popularity [61].

In 2015 I. Bali et al conducted a comparative analysis between the Karydakis and Limberg techniques, in which the researches have noted the Limberg technique to be advantageous according to all parameters: the duration of hospital stay – 1.4 against 3 days, early regaining of mobility by the patient – 1 against 2 days post-surgery, return to normal activities – 8 against 17 days, duration of overall healing time – 22 against 24 days. Finally, it was found that the Limberg technique resulted in higher overall patient satisfaction [40]. However, M. Ates et al (2011), in their research have noted a lower rate of recurrence in patients treated with the Karydakis method, as opposed to the Limberg method [41].

According to many modern authors, surgical techniques completed by lateralization of the wound show better results compared to other surgical approaches, in which lateralization is not used [41].

As to the closed surgical techniques and their modifications, researchers list the inactivation, of the trichogenic pump, due to the reduction in depth, or the complete flattening of the intergluteal cleft. This results in the reduction of skin debris present, normally present in the cleft, and reduces the possibility of hairs infiltrating the wound. The residual cavity is also less likely to form [45].

J. Bascom, as a proponent of the acquired theory of PD pathogenesis, proposed a number of surgical techniques in the 1980ies. One such technique is called Bascom I, and it is based on subdermal removal of the pilonidal cyst, beginning at the primary opening of the pilonidal fistula, which is later sutured, and finishing at the secondary opening, through which drainage later occurs. These measures reduced pressure within the postoperative wound, and allowed Bascom to achieve an average postoperative healing time of 39 days. 68% of the wounds were noted to heal by primary intention, with recurrence of 7.3% [23].

Some time later, J. Bascom developed a new surgical approach, called Bascom II (cleft lift), according to which, after the subdermal excision of the cyst from the intergluteal cleft, a thick skin flap is formed, which is place eccentrically, thus closing the residual openings within the cleft. At the end of the procedure, the intergluteal fold becomes "smoothed out", in other words maximally flattened. The Bascom II procedure has been demonstrated to be especially effective during repeated surgeries [20,23].

According to the majority of modern researchers, shaving, or laser removal of hairs surrounding the postoperative wound until the time of its full healing is crucial to the success of surgical intervention aimed at treating PD. Researchers also agree that maintenance of through personal hygiene around the postoperative area is very important [48,49,50,51].

Another important aspect of surgical intervention is a thorough and precise alignment of the postoperative wound edges. If the edges are not well aligned, a so-called "false recurrence" of PD may occur, characterized by uneven healing, and formation of a residual cavity underneath the scar, with subsequent penetration of hair inside the said cavity [52,53].
C. Soll (2001) proposed a minimally invasive method of PD treatment, characterized by subdermal resection of the primary and secondary fistulas, followed by the formation of a tunnel in the subcutaneous fat tissue, which connects the proximal and distal openings. According to the author, notable advantages of this method are in the formation of a relatively small residual cavity, a low likelihood of deformation of the sacrococcygeal region, a low rate of recurrence (7%), minimal postoperative pain, and a short healing time (under 10 days) [47].

P. Meinerro (2013) proposed a technique of endoscopic pilonidal sinus treatment (EPSIT) characterized by the destruction of coccygeal passages by a monopolar coagulator, with assistance of a fistuloscope, inserted through the secondary opening. This technique allows for the removal of hair, hair follicles, and various other debris from within the fistula [63]. A. Mortiz et al (1997) proposed treating PD with a carbon dioxide laser [59].

I.M. Nordon et al (2009) conducted a study, comparing the outcomes of Bascom I and Bascom II techniques. According to the results, the average healing time following Bascom I procedure was 4-4.5 weeks, while the average healing time after Bascom II procedure was 10-13 days [46].

M.H Abo-Ryia et al (2017) published the results of treatment according to a modified Karydakis procedure. During the surgery, the authors mobilized a fascial-dermal-subcutaneous graft, which was subsequently attached to sacral fascia, resulting in a satisfactory outcome in 96.7% of the patients [47].

In 2018 a team of researchers submitted a meta-analysis, in the form of a general evaluation of data, in which they gathered results from a total of 101313 studies by various authors, published from 1833 to 2017. From the gathered data, the team or researchers concluded that Karydakies, Bascom, Limberg and Dufourmentel procedures, as well as their modified variants show the least amount of recurrence [54].

CONCLUSIONS

In summary, it can be concluded that PD is an important socio-economic issue, as the patients are most often work-able adults (16-25 years of age). However, existing methods of treatment don't entirely satisfy neither the physicians, nor their patients, due to a high incidence of less than favorable outcomes. Furthermore, despite the extensive research, there still doesn't exist a universal consensus as to the etiology of PD and its definitive pathogenesis.

REFERENCES


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