Review



Labial Fusion in Childhood: Management and Treatment Strategies

Pediatrik Çağda Labial Füzyon: Yaklaşım ve Tedavi Seçenekleri

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ABSTRACT

Labial fusion (LF) is frequently an asymptomatic condition and hypoestrogenism plays an important role in the pathopysiology. In the presence of a hypoestrogenic environment friction i.e. in the form of vigorous perineal cleaning leads to inflamation and desquamation of the epithelium which results in LF. Expectant management and reassurance of the family is key in asymptomatic patient. However if the condition results in complications such as urinary retention, recurrent urinary tract infection or vaginitis topical therapy with estrogen or bethametazone is the first treatment option. For patients who do not respond to a course of medical treatment or for cases in which recurrence occurs manuel or surgical seperation may be offered. Regardless of the choice of treatment the family should be counselled of the possibility of recurrence. Recurrence rates decrease with increasing age and the commence of endogenous estrogen production. Factors that contribute to recurrence are poor genital hygiene, recurrent vulvovaginitis and vulvar dermatoses.

Keywords: Labial fusion, labial adhesion, labial synachiae, labial agglutination

ÖZ

Labial füzyon (LF) çoğunlukla asemptomatiktir ve herhangi bir klinik şikayete yol açmamaktadır. LF patofizyolojisinde "hipoöstrojenemi"nin önemli rol oynadığı düşünülmüştür. Hipoöstrojenize ortamda sık ve aşındırırcasına yapılan perine temizliği lokal inflamasyon, epitelin deskuamasyonu ve doku iyileşmesi sırasında füzyonla sonuçlanmaktadır. Asemptomatik olan hastalarda herhangi bir müdahale gerekmemekle beraber aileyi telarş zamanı geldiğinde kanda östrojen miktarının yükselmesiyle beraber LF'nun kendiliğinden çözüleceği konusunda rahatlatmak yeterlidir. Ancak enflamasyon bulguları, vajinit, idrar yolu enfeksiyonu gelişirse geleneksel olarak kullanılan ilk tedavi yaklaşımı krem şeklinde topikal düşük doz östrojen tedavisidir. Topikal betametazon tedavisi de etkin bir alternatiftir. Medikal tedavi ile LF gerilemezse veya tekrar eden uygulamalarda rekürrens gelişirse labiumların manuel seperasyonu veya keskin insizyonla cerrahi ayrıştırılması gerekebilir. Tedavide hangi yöntem kullanılırsa kullanılsın aileye rekürrens olasılığı hakkında bilgi verilmelidir. Rekürrens riski artan yaşla ve endojen östrojen üretiminin artmasıyla azalır. Rekürrense katkıda bulunan faktörler arasında kötü genital hijyen, tekrarlayan vulvovajinitler ve vulvar dermatozlar yer almaktadır.

Anahtar Kelimeler: Labial füzyon, labial adezyon, labial sineşi, labial aglütinasyon

Introduction

Labial fusion (LF) or as it is also called in the literature, "labial adhesion", "labial synechia", "labial agglutination" is the state of the labia minora sticking together on the vestibule in the midline (Figure 1). In the area where there is adhesion, there is a whitegray translucent and fibrotic tissue with varying degrees of fibrosis called "raphe". The presence of raphe allows differential diagnosis between congenital anomalies and LF. In a study conducted in our country in 2020, Huseynov and Hakalmaz (1) classified LF

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according to the degree of fibrosis. In type 1 LF, the fusion tissue is translucent and very thin. In type 2 LF, the fused tissue is fibrotic and thick. The degree of fusion may vary from partial fusion in the anterior or posterior to complete fusion. Partial adhesions mostly occur posteriorly but can close the vaginal introitus. Although the exact etiology of LF is not fully known, various risk factors related to its development will be discussed below. LF is most frequently observed in premenarche and mostly between the ages of 2-7 (2). According to an epidemiological study conducted in the USA, the age distribution in which it is observed is given as 6-24 months (3). As diaper use decreases and the physical ambulation of the child increases, the probability of LF decreases (4). The incidence of LF in girls in childhood is stated as 0.6-5% in different sources (5,6).

LF is mostly asymptomatic and does not cause any clinical complaints. In symptomatic cases, since it prevents urine flow in infants or prepubertal girls, complaints related to urination occur. Difficulty in urination, post-urination dripping or frequently recurring urinary tract infections are the most common symptoms we encounter in symptomatic patients. In addition, vaginitis may develop when synechiae close the vaginal opening and prevent vaginal secretions from draining. According to the study published by Mayoglou et al. (2) in 2009, 50% of the patients with LF were asymptomatic, while 19.9% had urinary tract infection, 12.6% had post-micturition drip, 8.6% had vaginitis, and 7.3% had frequent urination.

Risk Factors for the Development of Labial Fusion

It has been thought that "hypoestrogenemia" plays an important role in the pathophysiology of LF, which occurs with low estrogen levels at the two extremes of a woman's life; prepuberty and postmenopause. Since maternal estrogen, which passes from mother to fetus, persists in the newborn, LF is not



Figure 1. A partial labial fusion patient accompanied by vulvovaginitis. The patient was 20 months old and was brought to the hospital after being noticed by her family

observed in the first 3 months postnatally. In the literature, it has been determined that among the etiological risk factors, in addition to hypoestrogenism, perineal cleaning is also performed excessively by families (7). Frequent and abrasive perineal cleaning in a hypoestrogenized environment results in local inflammation, desquamation of the epithelium and fusion during tissue healing (8,9). Possible factors that may lead to local inflammation are given in Table 1. As a result of tissue trauma following infection or inflammation, the labial epithelium may erode and fuse. Simultaneous fecal contamination may also contribute to inflammation and result in vulvovaginitis. Some chronic inflammatory diseases, especially lichen sclerosus, have been associated with LF (10,11). Patients with poorly controlled diabetes, especially those with a history of recent antibiotic use, who develop candida vulvovaginitis, or infants who have frequent diaper dermatitis are also at risk for developing LF. Other infectious agents that may cause LF, although not very common, include N. gonorrhae, C. trochomatis, and T. vaginalis. It should be kept in mind here that if there is a history of sexually transmitted diseases in this age group, sexual abuse should definitely be excluded.

Recommended Clinical Examination for Labial Fusion

If the patient is under 8 years of age, it is a rational approach to examine her in the mother's lap with the legs in the "butterfly" position (12). This approach is a basic principle in pediatric gynecology, especially in terms of gaining the trust of young children and families who have received training that strangers should not touch the genital area. After being positioned in the supine position on the mother's lap or with the hip joint abducted and the heels touching each other, the examination consent is obtained with a calm voice. The urethral meatus and vaginal intrauterine occlusion should normally be visualized with gentle posterolateral traction of the vulva. Differential diagnosis of adhesions covering the entrance to the vaginal orifice, imperforate hymen and vaginal agenesis should be made. If this

| Table 1. Etiological risk factors that may lead to labial fusion |
|---|
| Poor genital hygiene |
| Trauma |
| Excessive perineal cleaning |
| Exual abuse |
| Female circumcision |
| Infection |
| Candida |
| Group A streptococcus |
| Gardnerella vaginalis |
| Chlamydia trachomatis |
| Trichomonas vaginalis |
| Vulvar dermatoses |
| Lichen sclerosus |
| Diaper dermatitis |

distinction cannot be made at the first visit, the vaginal orifice should definitely be inspected with a thin cotton-tipped swab at the post-treatment control visits.

First-stage Approach in Treatment

Although no intervention is required in asymptomatic patients, the applied approach can be expressed as "wait and see". It is sufficient to reassure the family that LF will resolve spontaneously with the increase in the amount of estrogen in the blood when thelarche time comes. Indeed, 80% of LF cases resolve without any treatment (13). In the presence of serious anatomical concerns, in cases of recurrent febrile urinary tract infection or vaginitis due to failure to drain vaginal secretions, the first-stage treatment methods are either topical estrogen-containing or topical betamethasone-containing preparations.

Topical Estrogen Treatment

Although most cases are asymptomatic and will not require treatment other than the "wait and see" approach, if signs of inflammation, vaginitis, or urinary tract infection develop, the traditional first-stage treatment approach is topical low-dose estrogen treatment in the form of a cream. Estrogen treatment protocols are usually applied as a thin layer to the fused area once a day for 4-8 weeks (14-17). Although thinner and more transparent adhesions are more likely to open, a success rate of 50-89% has been reported after estrogen treatment (18). No direct relationship has been reported between the duration of treatment and the resolution of labial adhesions. Recurrence rates ranging from 11-41% have been reported with this method (2,17,19-21). It is also necessary to mention the theoretical risks of long-term estrogen treatment. Increased labial blood flow and labial hyperpigmentation may be observed during treatment (22). Premature breast budding or breast tenderness may be observed as side effects in higher dose applications.

Topical Steroid Therapy

Topical betamethasone (0.05-0.01% concentration) treatment is the most commonly preferred agent (5,23). Betamethasone treatment was first used successfully in the treatment of phimosis in boys (24). With application twice daily with light traction, 67-95% of cases were successfully treated (25). In one study, in cases who were treated primarily with estrogen cream and then had recurrence, 68% of LF was reported to open after 4-6 weeks of twice daily betamethasone treatment (14). Betamethasone side effects include erythema, itching, folliculitis, skin atrophy or growth of fine villi hairs in the application area (26).

Approach in Cases Resistant to Medical Treatment

If LF does not regress with medical treatment or recurrence develops with repeated applications, manual separation of the labia or surgical separation with a sharp incision may be required. In addition, if there is a serious complication such as urinary retention, manual separation under sedation/local anesthesia or surgical separation is indicated. Among the techniques reported in the literature, there are studies in which manual separation is performed with the help of local sedatives and/or anxiolytics in

the office environment (27-29). Local anesthetic creams with a combination of 2.5% prilocaine and 2.5% lidocaine are suitable for use in pediatric patients. Topical anesthetics can be used in surgical separation, and sedation can also be given in the operating room (30). Topical estrogen or betamethasone preparations have been described to be used during or after separation techniques (17,29). It is recommended that topical treatments be continued for 2-4 weeks to prevent reepithelialization after surgical treatment, followed by the use of vaseline-like preparations to moisturize the perineum for an additional 3-6 weeks to reduce recurrence (5,17). As with any treatment, compliance and maintenance of genital hygiene are essential after surgical treatment. After the surgical incision, the healing epithelium should be prevented from adhering to the midline again. For this purpose, creams containing antimicrobial agents such as local vaseline or polymyxin were applied after the procedure (30).

Recurrence

Regardless of the method used in treatment, the family should be informed about the possibility of recurrence. The risk of recurrence decreases with increasing age and increased endogenous estrogen production. Factors contributing to recurrence include poor genital hygiene, recurrent vulvovaginitis, and vulvar dermatoses. The method used in primary treatment does not determine the possibility of recurrence (18). When recurrence occurs, treatment approaches are the same as in the primary case. Observation without treatment can be done, topical treatment can be given again, or surgery can be tried. There are not many studies in the literature on recurrent LF cases. According to one study, repeated use of topical estrogen in recurrent cases was found to be effective in 35% of cases (21).

Footnotes

Authorship Contributions

Surgical and Medical Practices: A.F.G.K., Ç.Ç., Concept: A.F.G.K., Ç.Ç., Design: A.F.G.K., Ç.Ç., Data Collection or Processing: A.F.G.K., Ç.Ç., Analysis or Interpretation: A.F.G.K., Ç.Ç., Literature Search: A.F.G.K., Ç.Ç., Writing: A.F.G.K., Ç.Ç.

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