Review Fused labia: a paediatric approach

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Key content:
• Labial fusion is common among prepubertal girls.
• In the majority of cases it is asymptomatic and should not be treated.
• In symptomatic children, initial treatment with local estrogen is advised. Surgical treatment should be avoided if at all possible and considered only as a last resort.
• Recurrence rates are high. The condition resolves with the onset of puberty.

Learning objectives:
• To learn how to diagnose labial adhesions.
• To be able to reassure parents that the condition is self-limiting and benign.
• To know the treatment modalities available for symptomatic children.

Ethical issues:
• There is no evidence that child sexual abuse causes labial adhesions, however, if the presentation is atypical or there are other concerns, advice should be sought from the lead professional for child protection in the organisation or Trust.

Keywords adhesions / labial agglutination / lichen sclerosus / local estrogen treatment / vulvovaginitis
Introduction
Fused labia (Figure 1), also known as labial agglutination or labial adhesions, is a common condition of the prepubertal girl and, along with vulvovaginitis, it constitutes one of the most common complaints presenting to paediatric gynaecologists.

Epidemiology
The prevalence of fused labia ranges from 1.8–3.3%, depending on the age of the children assessed, but accurate estimates are difficult as a proportion of children are asymptomatic and the condition may remain undetected. This was demonstrated in a study that aimed to set standards of normal genital anatomy among prepubertal children, where labial adhesions were found in 38.9% of girls. This included adhesions as small as 2 mm in length that were visible after colposcopic magnification.

Fused labia occur when estrogen levels are low and are therefore extremely rare in the first 3 months of life when maternal estrogens are still abundant in the infant's circulation. In a study of the genitalia of 9070 newborns, none had labial fusion at birth. The condition is self-limiting and corrects itself naturally in early puberty when the endogenous production of estrogen starts. The peak incidence is between 2 and 4 years of age. This also corresponds with the peak incidence of vulvovaginitis, which may coexist with labial fusion.

Predisposing factors
The skin of the vulva prior to puberty is fragile and thin and, as a result, is prone to infection and inflammation. Vulvovaginitis, diarrhoea and scratching of the vulva can lead to local trauma of the skin with superficial denudement of the labia. During the healing process, the edges of the labia fuse due to their close proximity and a thin membrane forms, starting at the level of the posterior fourchette and extending anteriorly, obliterating part or all of the vestibule.

In some children labial fusion is associated with nappy rash and may resolve once the child is toilet trained. Labial adhesions have been associated with lichen sclerosus, in which adhesions are characteristically dense and difficult to treat and can persist into puberty and adulthood. Lichen sclerosus can lead to the formation of adhesions that involve the clitoral prepuce and can cause clitoral entrapment and neurovascular bundle compression with severe pain.

Unusual infections such as severe herpes and graft versus host disease have also been associated with labial adhesions and can occur in older patients.

In the past there has been debate as to whether labial adhesions can be caused by sexual abuse. A few case reports from more than 20 years ago suggested this, but it has not been demonstrated in any more recent studies. However, both adhesions and child sexual abuse are common and may co-exist, but it is not now thought that the two are linked.

Clinical presentation
(See Box 1.) Labial adhesions are, in the vast majority of cases, asymptomatic. Rarely, however, they can be associated with urinary symptoms, vulvovaginitis and pain.

Urinary symptoms occur as urine accumulates behind the adhesions, leading to an altered urinary stream or dribbling after micturition. Stasis can also predispose to asymptomatic bacteriuria or overt urinary tract infection. Urinary tract obstruction is rare, although cases of hydronephrosis that resolve following lysis of labial adhesions have been described in the literature. Occasionally, an altered urinary stream interferes with toilet training or leads to soiling during micturition.

Vulvovaginitis can be perpetuated by labial adhesions as vaginal discharge and debris accumulate behind the adhesions.

Pain is present in approximately one-quarter of children with labial adhesions who are referred to a doctor. This is manifested by complaints or avoidance of activities that involve straddle positions, pain during walking or pulling at nappies or underwear.
Management

(See Figure 2). Diagnosis of the condition is made by inspection of the vulva alone. Unlike in cases of ambiguous genitalia, the clitoris and labia majora are normal and a thin avascular membrane is clearly visible between the labia minora. The adhesions usually involve the posterior part of the vestibule, starting from the posterior fourchette and extending anteriorly towards the level of the urethra.

Social pressure and anxiety about the normality of the genitalia and the presence of the vagina are usually the driving forces behind bringing the child to the doctor. In these cases reassurance that the condition is benign and self-limiting is all that is needed. In asymptomatic cases, treatment is not required. The labial adhesions will resolve naturally when endogenous estrogen production starts, usually before the age of 9 years. There are no associated sequelae to the girl’s future reproductive and sexual life and the condition needs to be clearly differentiated in the parents’ minds from other conditions that can lead to ambiguity or abnormality of the genitalia.

Irrespective of whether labial fusion is symptomatic or not, advice on meticulous hygiene of the vulva is recommended, as for vulvovaginitis. Regular cleaning is important, especially after defecation and urination. Avoiding local irritants, such as bubble bath, and choosing cotton rather than synthetic underwear is also advisable. Where vulvovaginitis coexists, a swab should be taken from the introitus and, if a pathogen is isolated, appropriate antibiotic treatment should be instituted.

Although labial adhesions are not thought to be caused by sexual abuse, both may occur at the same time. A clinician who has received appropriate child protection training should be responsible for assessment of any genital symptoms in a child. If the presentation is atypical or there are other worrying features during the consultation, advice must be sought from the lead professional for child protection with the organisation or Trust where the child is being seen. It must be remembered that the majority of children who are abused do not have any physical complaints related to trauma or infection.

If the adhesions are associated with a skin condition, such as lichen sclerosus, this is usually symptomatic and will need treatment. Lichen sclerosus leads to a typical appearance of the vulva, with parchment-like areas and foci of inflammation that look like ‘blood blisters’. Initial treatment will usually include local potent steroid cream application. Histological confirmation may be necessary if resolution does not occur or the appearances are atypical.

Topical treatment

In cases where labial adhesions are symptomatic and cause pain or urinary symptoms, treatment will become necessary. A conservative approach is usually suggested, consisting of local application of estrogen cream on the labia for no longer than 6 weeks. A pea-sized amount of estrogen cream (such as estriol 0.1%) is applied using a cotton bud along the line of fusion. Parents need to be warned against longer or excessive use of estrogen cream, as a proportion of the hormone is absorbed into the circulation, leading to systemic effects such as breast budding, uterine bleeding and local skin discoloration or erythema. All of these effects regress promptly after discontinuation of the medication. Success rates range between 50–100%, depending mostly on the density of the adhesions, the duration of estrogen application and whether or not there has been prior treatment and recurrence.

Based on observations on male children with phimosis, where local application of steroids can lead to resolution of the problem, betamethasone cream has been used in girls with labial adhesions. In a retrospective review of 19 girls, the majority of whom had previously undergone treatment of labial adhesions with either estrogen cream or surgical lysis, a course of local betamethasone cream 0.05% for 4–6 weeks was shown to be successful in 68% of cases. Nevertheless, in almost
one-quarter of cases the condition recurred. Although no adverse effects were reported in this study, betamethasone is a potent steroid that can be associated with local skin weakening and irritation, as well as potential concerns from systemic absorption. It is therefore not a recommended management option.

**Surgical treatment**

Where medical treatment has failed, and if there are associated urinary symptoms, surgical correction may be indicated. Whilst there are many reports of adhesion separation in the outpatient setting, this procedure, albeit brief, can be painful and traumatising and is not common practice in the UK. According to a survey of parents whose children had undergone separation of labial adhesions under local anaesthetic, the perception of their child’s discomfort was rated as moderate or severe in half of the cases.¹⁰ The success of separation in the outpatient setting has been reported at the rate of 80%,¹¹ but this needs to be balanced against the distress caused to the child.

In those few cases where surgical treatment is required, this should be carried out under a brief general anaesthetic. The adhesions are divided by using gentle labial traction or by running a sound along the line of fusion. There is usually no or minimal blood loss and suturing is not usually necessary.

Recurrence is a common problem in labial adhesions after both application of estrogen cream and surgical separation. Recurrence rates range between 10–16%, depending on the treatment.¹² These numbers are probably underestimations, as most published studies have short follow-up periods. Recurring adhesions, particularly where there has been prior mechanical separation, are denser and therefore less likely to resolve with conservative management.¹³ There is also some suggestion that, following prior surgical separation, recurring adhesions may be less likely to resolve with the onset of spontaneous puberty. In the majority of these cases repeat surgical separation will be required. This reinforces the commonsense approach of only treating adhesions when they are symptomatic, as treatment can be futile and can lead to the formation of stronger adhesions after some months.

**Conclusion**

Labial fusion is a relatively common condition that can cause significant distress to parents because of worries regarding the normality of their daughter’s genitalia. Doctors must be firm that the condition is benign and resist any temptation to treat the condition unless the child is clearly symptomatic. Recurrence is common and all treatment options have potential physical or psychological adverse effects. There are no large randomised trials on management of labial adhesions that clearly clarify the ideal treatment and most treatment algorithms are based on the doctor’s experience or preference.¹⁴ However, it is logical to start treatment with the less invasive method of local estrogen application and only proceed to surgical separation under general anaesthesia when conservative treatment has failed.

**References**