

GENITAL OEDEMA

Justine Whitaker

Chronic oedema of the external genitalia has been classified as congenital or acquired (McDougal, 2003), and, as with other oedemas, is often referred to as 'primary' or 'secondary' lymphoedema.

Congenital or primary genital oedema is either infrequent in presentation (acquired during fetal development) or inherited. Acquired or secondary genital oedema can be caused by:

- ▶ Cancer and/or its treatment
- ▶ Infection (cellulitis, lymphangitis)
- ▶ Granulomas, e.g. in Crohn's disease
- ▶ Idiopathic causes.

Scrotal filarial infestation is also responsible for male genital oedema in developing countries (Garaffa et al, 2008). External female genitalia can also be affected. Thorough assessment, coupled with an understanding of the anatomy of the pelvic lymphatics will assist in a comprehensive differential diagnosis (Tiemstra and Kapoor, 2008).

When the scrotum becomes oedematous, it is important to decide whether the condition is acute or chronic to rule out potential medical emergency, such as testicular torsion. Pain and sudden swelling is usually associated with acute onset. Colour flow Doppler imaging has been shown to be an accurate diagnostic test in

identifying testicular torsion, for which surgery is the only treatment option (Burks et al, 1990). Failure to treat the torsion could result in permanent damage and, at its worse, necrosis and loss of testes.

Weinberger et al (2007) formulated an algorithm for the differential diagnosis of scrotal oedema to improve treatment planning. This algorithm highlights the need for multidisciplinary working to achieve the best outcome for patients. However, it mainly covers acute oedema, with chronic oedema forming only a small part of the pathway. *Figure 1* redresses this balance, demonstrating how chronic oedema of the scrotum deserves its own place in the management process.

Soft pitting tissue, which is generally painless, is the first sign of genital oedema, becoming chronic when it has been present for three months or more. In time, as with chronic oedema of the limbs, the tissue becomes fibrotic, brawny, misshapen and skin changes occur, such as hyperkeratosis and papillomatosis. Lymphangiomas (also known as lymphangiectases, depending on the depth of tissue) appear in the genital area. These have a wart-like appearance and are often mistaken for sexually transmitted genital warts (*Figure 2*). The lymphangiomas can be few or several in number. They generally manifest as small translucent vesicles (or papules), maturing into lymphangiomas. In both primary and secondary lymphoedema, during the translucent vesicle and lymphangioma stage, lymphorrhoea may leak either constantly or intermittently.

Treatment options

Both the male and female genitalia can grow to monstrous proportions once oedema becomes established. The scrotum contains no bony or muscular structures to allow/enhance lymphatic return and, therefore, can be considered as a 'dependent organ' when considering treatment options.

The last decade has seen an increase in lymphoedema services, which offer conservative treatments. Before this, surgery was the main treatment option. However, long-term results of surgery have not always proved beneficial. Browse et al (2003) obtained results over 10 years and found that only around half of patients had their symptoms adequately improved, the other half had to have second surgical procedures five to seven years later to control the recurrence of oedema. In addition, Osborne et al (2000) highlighted that until the recent advances in magnetic resonance imaging (MRI) contrast scanning, lymphangiography was the only option to determine the extent of deep lymphatic involvement before surgery (should this be the treatment of choice). This procedure, which can be technically difficult, also carries the risks of invasive procedures.

If simple surgical excision were to have been undertaken of the visible lesions, as was normal in this presentation, this would not have been curative (Osborne et al, 2000). Laser therapy has also been used in the treatment of genital oedema. Egan et al (1998) showed how they managed multiple scrotal lymphangiomas by treating them with carbon dioxide

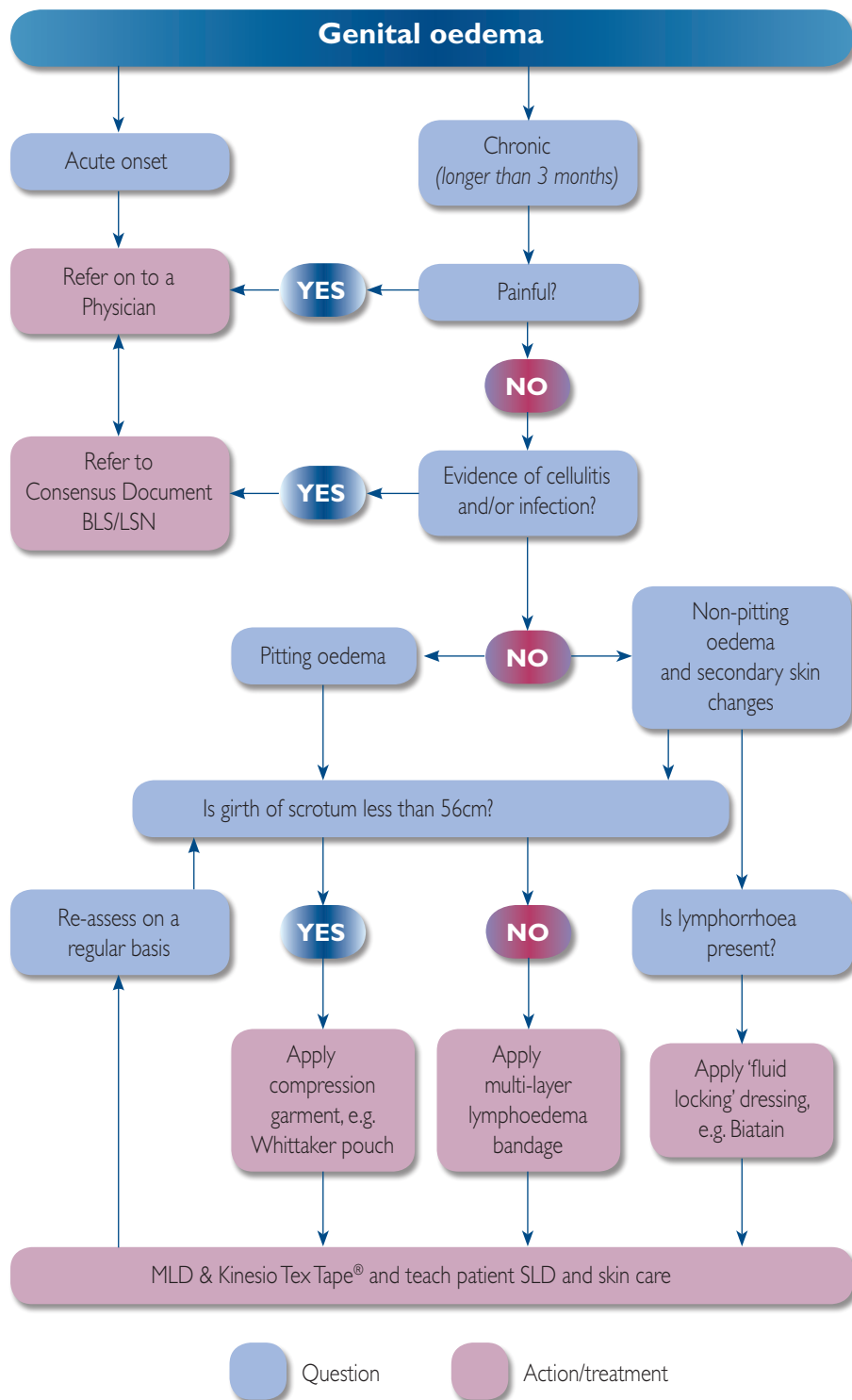


Figure 1. Whitaker genital oedema algorithm.

laser ablation. Anthony et al (2002) found that the Egan technique alone often resulted in a recurrence of the lymphangiomas. They took this technique and added cutting diathermy under anaesthesia which reduced the recurrence of lymphangiomas (Anthony et al, 2002).

Conservative treatment options are similar to those used for both upper and lower limb oedema.

Current literature supports the use of manual lymphatic drainage (MLD) to remove fluid from congested, damaged lymphatic tissue

to areas where the lymph flow is normal, thus reducing oedema (Harris and Pillar, 2003). The case reported in this article supports this theory. Teaching people affected with genital oedema or their partners/carers a modified version of MLD, simple lymphatic drainage (SLD), can help to maintain the reduction and softening of tissues that is achieved with MLD. It can also begin to address issues that individuals may have concerning their sexuality.

As with limb oedema, compression garments should be worn daily as a maintenance therapy after successful decongestion by SLD/MLD. This is particularly the case with male genital oedema where an elastic pouch can help.

Female genital oedema is more challenging in that there are currently no 'off-the-shelf' garments manufactured purely for managing oedema in the vulval area. Where specific garments and appliances are not readily available to certain patient groups, innovation and exploration in other fields can often produce solutions. For example, the 'V2 Supporter' (a prenatal cradle) which is available during pregnancy for vulval varicosities, in the author's experience and according to manufacturer's recommendations, offers controlled support to the labia, and tension can be adjusted to control swelling in lymphoedema. Several manufacturers also offer an array of different density foams which are useful in both male and female genital oedema. They can be tailored for the individual to be worn inside supportive underwear (Figures 3 and 4).

Bandaging of the scrotum and individual bandaging of the penis are options. This technique can be taught to the affected individual and/or their carer/partner. Gültig in 2005 pointed out that bandaging should begin with low level compression and only be increased according to individual tolerance and consent. Written consent should always be obtained and boys with primary genital



Figure 2: Lymphangiomas leaking lymphorrhoea.



Figure 4: Whitaker pouch in situ with ribbed foam around pubis area.

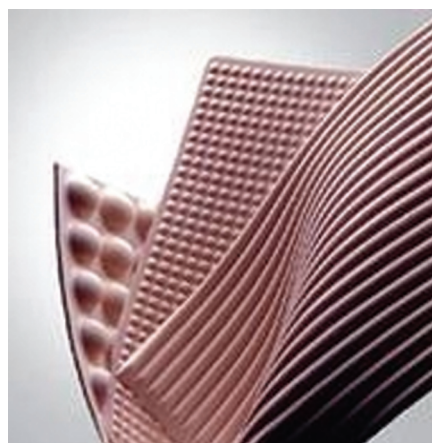


Figure 3: Foam accessories for use with compression.



Figure 5: Oedema on suprapubic area is rubbery, fibrosed and gross in size.

lymphoedema should be informed of the risk of fertility problems with long-term compression (Gültig, 2005).

Scrupulous hygiene and skin care are essential. As with limb oedema there is a risk of cellulitis, which is likely to be higher in people suffering from lymphangiomas. Daily washing and moisturising with a bland, lanolin-free cream should be encouraged. Although rare, the active ingredients in some emollients, triclosan and benzalkonium chloride, have been reported to cause irritant dermatitis of the genitalia (Saw and Hindmarsh, 2005).

The following case report gives an insight into how conservative therapy can play an important role in managing genital oedema.

Case report

A 64-year-old gentleman with gross idiopathic lymphoedema of his scrotum and supra-pubic region presented with a ten-month history of symptoms. There was no specific trigger for the oedema other than his scrotum became red and swelling appeared, associated with clinical symptoms of cellulitis. He was treated with several courses of oral antibiotics, however, the

symptoms persisted and he showed no signs of improvement.

His condition was exacerbated some six months after the initial onset by a holiday, where his ankles began to swell and his scrotum grew to monstrous proportions. Since this vacation he has been on long-term antibiotics of a daily dose of penicillin (2g).

At the initial consultation the oedema in his scrotum was pitting, but felt hard and tight, and the oedema in his supra-pubic area was rubbery,



Figure 6: How to measure the scrotum.



Figure 7: Six weeks into treatment, noting reduction in size of pubis and scrotum and softening of tissues.

fibrosed and gross in size (Figure 5). His penis lay flat within the scrotum and was undefined. As a result, he had problems with micturation. As seen in Figure 2, he had marked lymphangiomas which leaked lymphorrhoea. Due to the volume of fluid that leaked onto his underwear and trousers, which had

caused great embarrassment on many occasions, he wore a hand towel in his underwear at all times. This constant 'wetness' made him self-conscious of odour and potential skin breakdown. A lymphoscintigraphy was requested to establish the state of the pelvic lymphatics proximal to the problem, e.g.

hypoplasia/hyperplasia, or whether it was simply due to an acquired infection. However, due to the positive response to treatment after six weeks, the consultant dermatologist decided that this was not necessary and so the scan did not go ahead.

At the initial consultation the following measurements of the scrotal area were taken in a standing position with the scrotum dependent (Figure 6). This was to gauge the effectiveness of treatment and ensure that the correct size of garment was supplied:

- A: Girth of scrotum (widest part middle of scrotum) = 39cm
- B: Girth of neck of scrotum = 36cm
- C: Length of scrotum (base of penis to perineum) = 21cm

Treatment plan for initial 12 months included:

- ▶ Manual lymphatic drainage (MLD) to:
 - improve and enhance lymphatic pathways
 - reduce congestion in the affected area
 - help reduce fibrosis of the suprapubic region
 - to prevent the condition from worsening.
- This consisted of an initial course of MLD with a lymphoedema specialist twice a week for six weeks, once a week for four weeks, then once a fortnight for four weeks, followed by monthly 'top-up' sessions (approximately 24 sessions).
- ▶ Provision of appropriate compression garments on a regular basis, including four large-size scrotal supports (Whitaker Pouch) every 3–4 months, with reviewing as necessary.
- ▶ Additional density foam to wear inside the pouch to break down fibrosis and assist drainage in appropriate lymphatic pathways.
- ▶ Teaching the patient and patient's partner SLD to maintain MLD outcome.
- ▶ An introduction to appropriate regular exercise to improve function, mobility and posture.
- ▶ Skin care to improve skin condition and reduce the risk of cellulitis,

i.e. washing daily and applying appropriate emollients upwards towards the axilla area.

- ▶ Psychological support and advice to help the patient come to terms with a life-long chronic illness and to address sexuality needs, as appropriate with partner's involvement.
- ▶ General advice, education and support to encourage self-management.
- ▶ Kinesio tape to improve lymph drainage, maintain the effects of MLD and reduce fibrosis in the supra-pubic area.
- ▶ Teaching the patient and his partner how to apply Kinesio tape correctly imbetween the MLD treatment sessions.
- ▶ Prescribing various appropriate absorbent pads/dressings (e.g. Biatain™ [Coloplast], 10x10cm, bevelled edge) while undergoing treatment, to be worn inside pouch.
- ▶ Requesting radio-nuclear tests from consultant/GP.

Treatment outcome

At six weeks, a vast improvement could be seen in the patient's condition (i.e. after application of the pouch, daily skin care and daily SLD supported by MLD twice a week).

Psychologically, the patient showed positive progression and was planning a wedding later in the year. He was self-managing his condition and no longer required a hand towel in his underwear daily. A vast reduction in the amount of lymphorrhoea was noted and on most days the Biatain was completely dry. The pubic area was markedly softer and reduced in size (Figure 7). His scrotal measurements at this time were:

- A: Girth of scrotum (widest part mid way) 32cm, a reduction of 7cm
- B: Girth of neck of scrotum = 30cm, a reduction of 6cm
- C: Length of scrotum (base of penis to perineum) = 17cm, a reduction of 4cm

The pouch size was reduced from a large to a medium size, to maintain

compression and help further to reduce the oedema.

The infection appeared under control and asymptomatic, and the ankle oedema had resolved. He remains on antibiotics and is being reviewed by his dermatologist on a six-monthly basis.

Conclusion

A thorough understanding and knowledge of both the superficial and deep lymphatic system is required to ensure optimal treatment for the differential diagnosis of genital oedema. Patients' symptoms often prove to be idiopathic. With a systematic assessment procedure, genital oedema can be managed effectively by conservative therapy.

Discussion with other members of the multidisciplinary team is essential to rule out the possibility of an incorrect diagnosis. On rare occasions, surgical intervention may be necessary.

Providing the patient with the essential tools and package of care which they can carry out on a daily basis is the key to gaining long-term benefits. Addressing genital oedema in a robust way enhances the patient's well-being, both psychologically and clinically, enabling them to live a full and active life, which does not revolve around hospital appointments. **JL**

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Key points

- ▶ Genital oedema can be either primary or secondary.
- ▶ Thorough assessment, coupled with an understanding of the anatomy of the pelvic lymphatics will assist in a comprehensive differential diagnosis.
- ▶ Treatment options can be both conservative and surgical.
- ▶ Scrupulous hygiene and skin care are essential. As with limb oedema there is a risk of cellulitis, which is likely to be higher in people suffering from lymphangiomas.
- ▶ Measurable treatment outcomes can be achieved.

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