# Venous leg ulceration in a child secondary to an iatrogenic arteriovenous fistula: the role of compression therapy

KEY WORDS

- Venous leg ulceration
- Child
- Compression
- Iatrogenic arteriovenous fistula

The case study presents the account of a two-year-old female with a medical history of Mosaic Trisomy 9, congenital vertical talus and developmental delay who presented to the Tissue Viability team with venous leg ulceration and swelling to the right leg. The extent of the venous ulcer at the time of presentation put the limb at risk of primary amputation. The family have kindly consented to share the photos and journey of the leg. The illustrated narrative depicts the early stages of skin breakdown before joint Tissue Viability and Vascular assessment and care as a case study of effective multidisciplinary practice. Compression therapy was instigated as the first-line treatment; initially to the knee level and subsequently to the groin using an elasticated four-layer system. Preliminary review of the literature, discussions with experts and industry suggested the lack of evidence to support the role of compression to treat paediatric active venous leg ulceration. We aim to demonstrate the healing potential of compression therapy applied in a systematic and skilled way by a competent practitioner. This case, which was nurse-led and carried out over a five-month period, adds to the limited body of existing evidence around venous leg ulceration in children. The report highlights the skill and knowledge required to treat complex cases and encourages nurse practitioners to think outside the box and work together with our vascular surgical colleagues to achieve results.

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A two-year-old female who will be named 'Kelly' for anonymity was born with Mosaic Trisomy 9. According to the Oxford Concise Medical Dictionary (2014):

*'Mosaicism is a condition in which the cells of an individual do not all contain identical chromosomes; there may be two or more genetically different populations of cells. Often one of the cell populations is normal and the other carries a chromosome defect such as Down's syndrome or Turner's syndrome'*

Trisomy 9 is a rare chromosomal abnormality that can occur in a mosaic or non-mosaic state. Full trisomy 9 is rare in live born infants but trisomy 9 mosaicism has been reported to be compatible with life. Clinical manifestations in Kelly's case (*Figure 1*) included developmental delay, atrial septal defect and congenital vertical talus (Pejcic et al, 2018). At six months old Kelly underwent an atrial septal defect (ASD) closure.

At ten months of age Kelly underwent above-the-knee serial casting for two weeks and then closed reduction of talonavicular joint and percutaneous achilles tenotomies. Following this procedure, a cast was worn for a further six weeks. Two months later, now one year old, Kelly presented to the Emergency Department with increased swelling of the right lower limb (*Figure 2*). At this point, the skin was intact; however, early signs of chronic

Figure 1. At 6-month-old Kelly before atrial septal defect closure procedure.

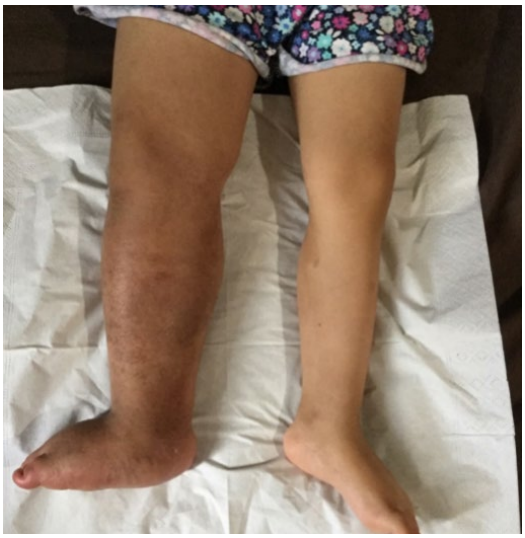

Figure 2. Kelly at one-year-old with swelling of the right lower limb

venous hypertension and insufficiency (CVI) with associated oedema and haemosiderin venous skin staining could be observed (Lurie et al, 2020). A total of three venous scans were undertaken at 12, 15 and 16 months; all interpreted as showing patency of the major veins in the leg.

## A timeline of events

At 15 months of age:

- Markedly increased swelling; admitted with below knee cellulitis; treated with intravenous antibiotics

At 16 months of age:

- Conflictual vascular findings
- MRI right limb suggesting extensive deep vein thrombus (DVT) in the external iliac vein distally and stenosis in the femoral vein at the level of the sapheno-femoral junction; commenced on low molecular weight heparin (LMWH)
- Venogram: no clot in deep or superficial system but very convoluted superficial veins with conclusion that it probably represents a congenital venous anomaly
- Discussed in multidisciplinary team (MDT) and referring hospital advised to stop LMWH and start compression therapy.

At 19 months of age:

- Seen by Dermatology in outpatient clinic and advised compression with possible local referral to lymphoedema services for measured compression garments.

At two years of age:

- Kelly had begun to develop a pre-ulceration on the right medial malleolus. At this point the child was not receiving compression therapy, had two admissions for cellulitis and received topical antimicrobial treatment to the area *(Figure 3)*. Within eight weeks from the onset of the pre ulceration observed at the right

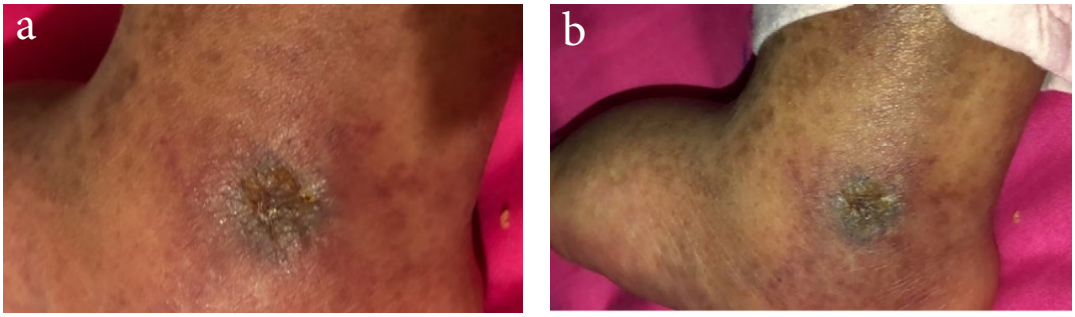


Figure 3. Onset of right medial malleolar ulceration

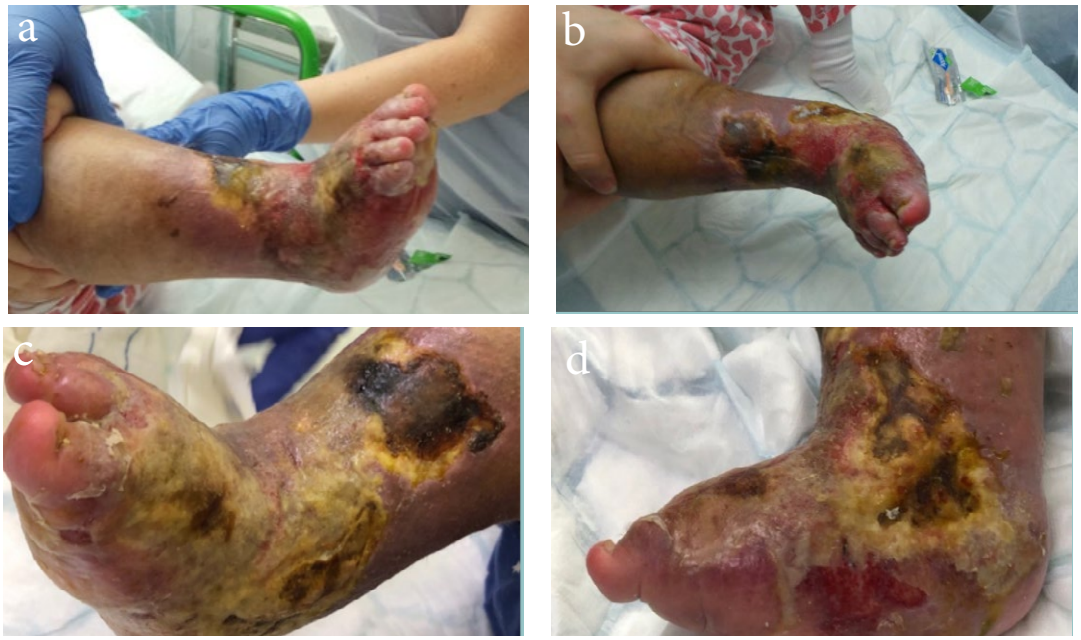


**Figure 4. Kelly, age two years and seven months old, when compression bandaging was started: Note the circumferential leg ulceration with extensive tissue loss and necrotic tissue**

medial malleolus, the skin had broken down and ulcerated.

Following referral to the tissue viability team at the Children's Hospital (RA) contacted their referring tissue viability nurse colleague (JG) to assess and instigate compression therapy; Kelly was 2 years and 7 months old when compression therapy was started.

As *Figures 4* shows, the leg ulceration was circumferential with extensive tissue loss and a significant amount of devitalised and necrotic tissue. The ulcer was producing heavy amounts of exudate, which was causing excoriation to the surrounding skin. There was significant swelling and pain from the ulcer and Kelly required analgesia before and after the dressing change. The dressing change itself was distressing for Kelly; Mum and Dad were always present and supported the nursing staff by comforting the child and holding the leg during wound assessment and dressing change. The time required to complete the dressing at this stage was 60–90 minutes every 2–3 days, depending on exudate levels. Antimicrobial dressings were used to keep the wound bed moist and provide topical antimicrobial treatment. Wound dressings, usually antimicrobial, were used under the direction and agreement of the paediatric team due to potential risks; these included topical iodine products with a betaine/polyhexanide solution to cleanse.

The paediatric tissue viability team (RA) at the Children's Hospital were not trained in compression bandaging therapy therefore this was instigated by JG with the initial intention of training the team to continue the therapy with regular support and weekly review at the Vascular Clinic. One of the measured outcomes was the full circumference of the lower limb at the level of the foot, ankle, calf, below knee, above knee and groin. Recording measurements were paramount to document the smallest reduction in oedema in the context of extensive ulceration.

Despite compression therapy and alternate day dressing changes, the right leg ulceration continued to deteriorate in the first few weeks of treatment; although the oedema was slowly reducing, there was no real progress and the child was not thriving. The parents also had to travel long distances daily to attend for dressing changes so there was limited rest for the family. It was decided the child would be admitted for daily compression therapy and rest. Following Vascular Consultant review, the limb was deemed borderline salvageable due to the extension of tissue loss and functional sequelae.

Despite the appearance *(Figure 5)* of the foot being borderline non-viable, the arterial perfusion

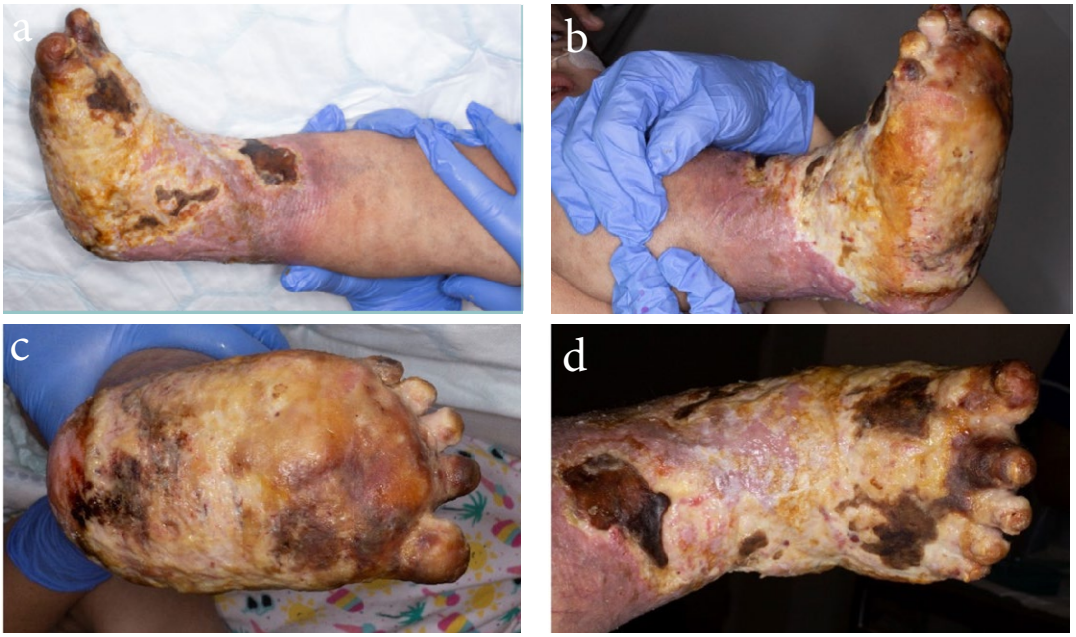


**Figure 5. Progressive deterioration in the first weeks of compression therapy; medial (a, b), plantar (c) and dorsal (d) aspects**

was preserved with strong, triphasic pedal signals on continuos-wave doppler.

The possibility of primary major amputation was explained to Mum, due to the risks of sepsis and further functional deterioration of the limb. Understandably, this caused the family a great deal of anxiety and, at first, confusion about what was happening. It was, however, agreed to continue with aggressive compression treatment of the full leg.

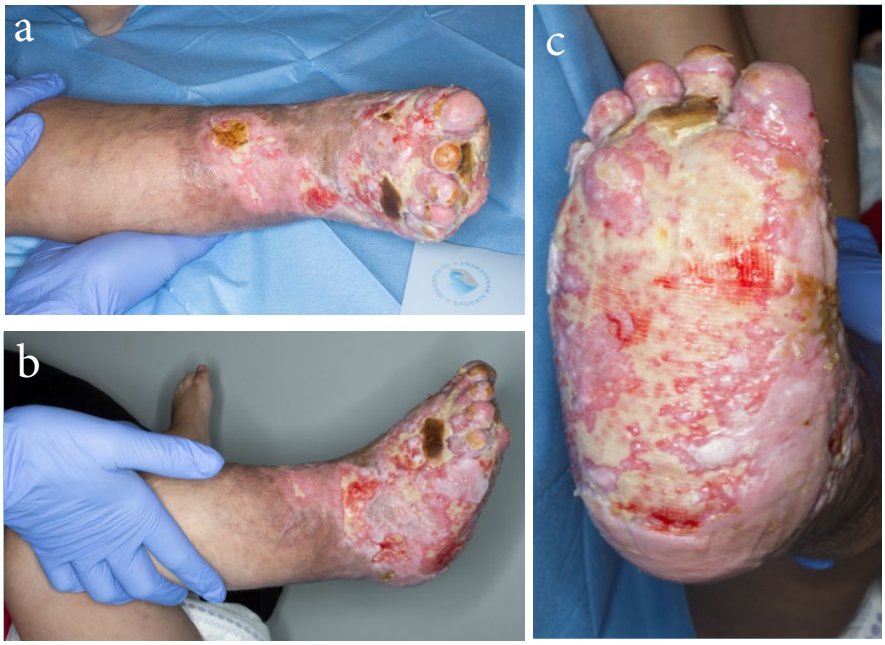


**Figure 6. Improvements following 3 months of compression therapy, bed-side sharp debridement and leg elevation. Healthy granulating tissue; dorsal (a), lateral (b) and plantar (c) aspects**

Dressing regime at this stage included betadine soaks, alongside washing the skin of the whole limb and sharp debridement of non-viable moveable tissue. Topical dressing included antimicrobials, superabsorbents and skin care with emollients. Compression was applied using the first three layers of the four-layer system applied at first in a spiral and then in a figure of eight, stretched at 50%. For the first four weeks this was to knee level only and then applied fully to the groin with the additional fourth layer, as agreed by the Vascular team

The strict care plan was repeated on a daily basis due to the heavy exudate from the wound, present even with the use of superabsorbent dressing. Kelly was hospitalised for five weeks and subsequently continued with outpatient daily dressings. At each dressing change, whether on the ward or in the clinic area, the dressing was complex as the child was often distressed and continuously moved. On average the dressing at this stage took over two hours

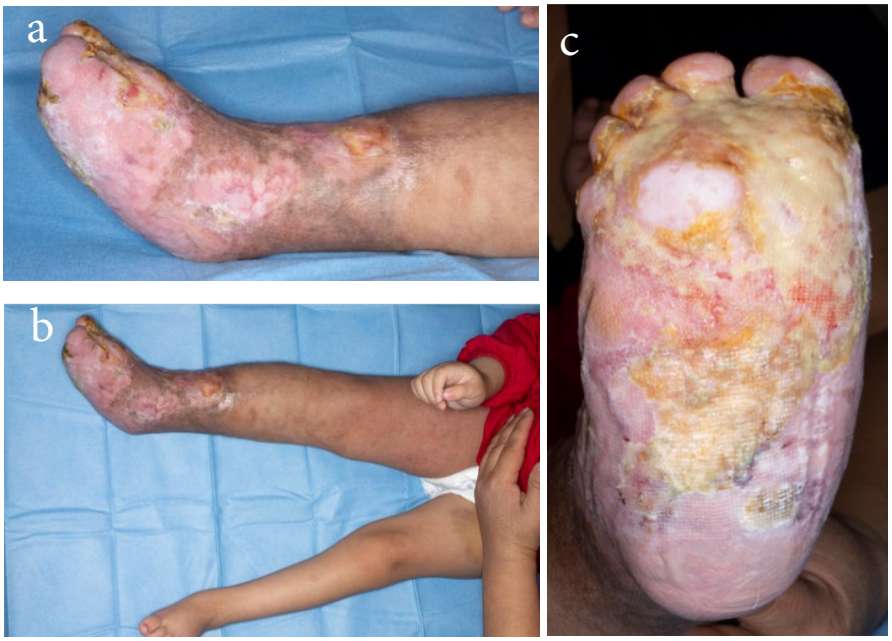


Figure 7. Kelly after deep vein angioplasty and ongoing compression bandaging

daily to apply, sometimes with two nurses if debridement was required. Sharp debridement was carried out by (JG) regularly and for the most part the child received oramporh before dressing change and was also on a monitored dose of Gabapentin. In the beginning, her mobililty was poor and she was often agitated, which meant she did not rest the leg. As her leg improved she naturally became more active. Overall elevation was feasible while sleeping; however, Mum did elevate the leg on a pillow when she travelled in her pushchair.

Following three months of regular compression therapy, the child began to thrive and gain weight; her hair started growing and she became physically and psychologically stronger: she started playing and dancing, her mood greatly improved and she was happy. This positively reflected on her mum.

At this stage *(Figure 6)*, further vascular investigation (venous duplex), identified an acquired/iatrogenic Arteriovenous fistula (AVF) and confirmed the initial diagnosis of supra (iliac vein) and infra-inguinal (common and femoral veins) deep vein thrombosis (DVT). AVFs are abnormal connections between the arterial and venous system that divert blood from the high pressure arterial system directly into the low pressure venous system (bypassing the capillary system), ultimately causing venous hypertension. Fistulas may occur anywhere in the body, be single or multiple and be congenital or acquired (Mylankal et al, 2011).

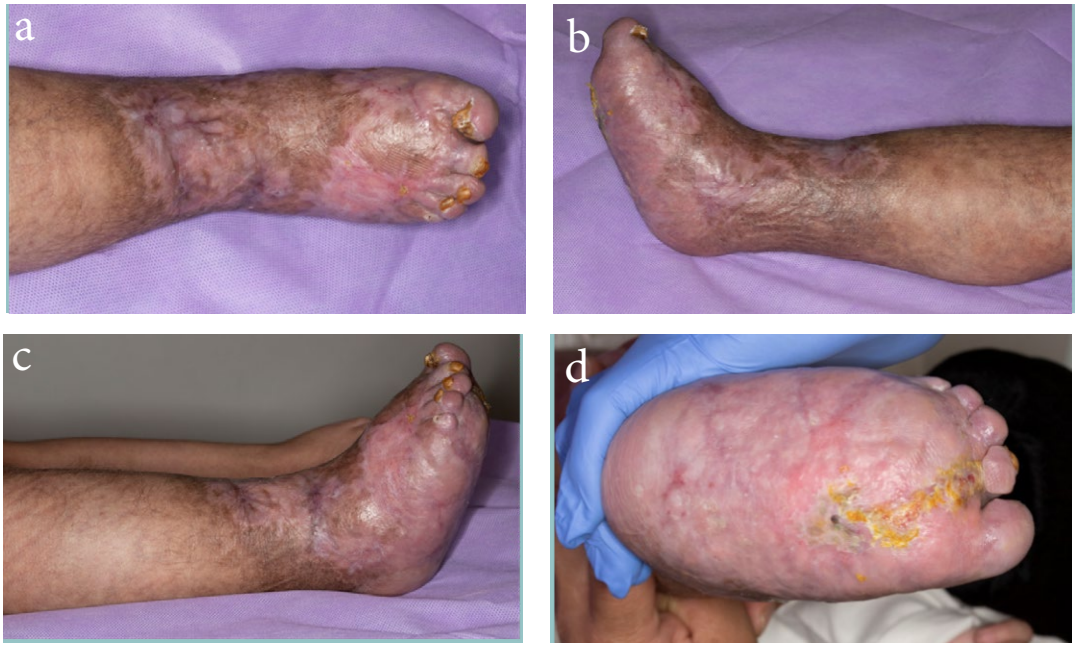


Figure 8. Full healing of the venous leg ulcer, following five months of compression therapy

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Aged two years and 10 months *(Figure 7)*, Kelly underwent an endovenous recanalisation of the iliac vein (deep venous angioplasty) to improve the venous outflow. The infrainguinal DVT and AVF remained untreated, with plans of considering the opportunity of surgical repair of the fistula at a later stage.

By the age of three the VLU had fully healed. The compression remained full four-layer to the level of the groin; the frequency of dressing was reduced as the leg ulcer began to improve from daily to alternate days and then three times a week, with particular attention to the compression bandage to the groin often rolling down the leg due to the activity of the child. In terms of the skin care topical regimen, emollient and non-adherent dressings and antimicrobials were used when required. Customised compression hosiery was not an option due to the severity of the ulceration and the frequency of dressing change; this was agreed by the teams.

As the final photos demonstrate *(Figure 8)*, five months after the initial review by tissue viability and commencing compression therapy, the wound had almost completely re-epithelialised with mild, superficial scar tissue. Although moderately oedematous and hyperthrophic (due to the longstanding AVF) the lower limb was viable, intact and functionally improved; Kelly was stronger, healthier and happier.

## CONCLUSION

This case report describes the successful multidisciplinary management of a complex venous leg ulceration in a young child and the key role of multicentre tissue viability care offering integrated adult and paediatric expertise. Everyone involved in the case worked for the child's benefit, across sites with no barriers of leadership, skills or role.

The case also embraces the complex nature of lower limb ulceration and the success of timely and appropriate compression therapy in paediatric patients, despite the lack of medical evidence, provided specialist multidisciplinary assessment, correct differential diagnosis and close clinical surveillance. WUK

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